Ipsilateral Lower Ureteroureterostomy for The Management of Ureteric Duplication Anomalies in Children: A Prospective Study

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Abstract

Introduction: Congenital Duplex anomaly of the ureters is a challenging clinical disorder, which is described in approximately 1 % of children with higher prevalence in girls. It arises from abnormal location of ureteric bud from wolffian duct during embryogenesis. A condition that can be diagnosed antenatally. The upper moiety usually ends with either a ureterocele or an ectopic ureter.

Management options include expectant conservative management, transurethral ureterocele incision (TUI), ipsilateral ureteroureterostomy, upper pole hemi nephrectomy and total reconstruction with common sheath reimplantation.

Evaluation of the short-term effect of lower ipsilateral ureteroureterostomy procedure for the management of ureteric duplication anomalies in children.

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Materials and Methods: A prospective study including thirty-two children with complete duplex anomalies in which the ipsilateral upper moiety ureter is affected from March 2023 to March 2024 were included in the study. All patients underwent open ipsilateral lower ureteroureterostomy.

Results: Thirty-two children (21 male, 11 female patients) underwent ipsilateral Ureteroureterostomy, 75% of the patients had ureterocele and the remaining had ectopic ureter. Mean age was 20.6 months.
Urinary tract Infection was the most common presenting symptom in 59.4%, Other presentations include antenatal hydronephrosis in 28.1% and incontinence in 12.5%.
Median follow up was 10.43 months (IQR:8.1-14.3). The percentage of children experiencing adverse events is 15.6%, in the form of post-operative UTI in 4 cases (12.5 %) and one case, in which the operator could not pass a JJ stent intraoperatively and performed subsequent reoperation.

Conclusion: Ipsilateral ureteroureterostomy has a low risk of surgical morbidities and low rate of complementary redo operation in children. We recommend performing it in duplex anomalies whose primary pathology is an ectopic ureter.

Keywords

- Ipsilateral lower ureteroureterostomy
- Duplex anomalies
- ectopic ureter
- ureterocele

Introduction

Ureteral duplication anomalies, which affect 1% of the population and are more common in females, are among the complicated congenital anomalies that pose a substantial clinical challenge for paediatric urologists. Ureterocele, and ectopic ureter are examples of upper moiety obstruction caused by ureteral duplications.[1]

Numerous genetic routes have been hypothesized, but the pathophysiology of ectopic ureters and ureteroceles remain unclear.[2] Aberrant ureterotrigonal development resulting in renal dysplasia might explain the pathophysiology of ectopic ureter and ureterocele.

According to Weigert Meyer rule, ureteroceles are often draining the upper moiety in duplex systems. Although there are several anatomical classifications for ureterocele, the most crucial clinical classification is the distinction between the intravesical and extravesical types.

Any ureter, with or without ureteric duplication, whose orifice is not at the typical location in the trigone is considered an ectopic ureter. The lower pole ureter is mostly introduced into the urogenital sinus. As per the Weigert Meyer rule, in a duplex system, the ectopic ureter mostly drains the upper pole.

Anomalies involving urinary duplications frequently show up after birth as lower urinary tract infections or pyelonephritis. They should always be considered in the differential diagnosis of recurrent post-natal urinary tract infections.[3] It is usual for girls with ectopic ureters to be wet most of the time. When a ureterocele prolapses in the neck of the bladder, it can obstruct the bladder neck and cause lowert tract obstructive symptoms.

It should be emphasised how crucial it is to obtain as much information as possible regarding the altered anatomy and physiology of the patient before undergoing any surgical intervention for ureteroceles or ectopic ureters. The therapeutic goals regardless the various techniques are the same: maintenance of urinary continence, clearance of infection, obstruction, and reflux, and preservation of renal function.[4] A goal that needs to be taken into account in this study is lowering surgical morbidity.

When conservative treatment is used on children without obstructing ureteroceles, up to 39% of them will eventually need surgery for recurrent infection. This method is recommended for children who have non-obstructing, small ureteroceles, low-grade vesicoureteral reflux, no active bacterial infection, and no obstruction of the bladder outlet. [4-5]

It has been found that 18% of intravesical ureteroceles require additional interventions in the future after transurethral endoscopic ureterocele incision (TUI), compared to 64% of ectopic ureteroceles (i.e., a higher reoperation rate for ectopic ureteroceles treated with TUI).

The effectiveness of TUI in treating duplex ureteroceles is controversial. Of the patients who underwent TUI for upper pole ureteroceles, 48% developed upper pole VUR, and 71% required a second open surgery.[6-11]

In situations with duplex system with minimal or non-functioning upper moiety; open or laparoscopic upper-pole partial nephrectomy is a very plausible choice; however, it may not resolve the problem entirely. Following hemi nephrectomy, clinicians record 20% of persistent reflux into lower-moiety or contralateral ureters. In addition to a regular finding of some amount of function loss.[12-15]

Ureteroureterosotmy (UU) requires a smaller inguinal incision and has a lesser risk of renal haemorrhage or damage to the ipsilateral lower pole, it is a less complicated surgery than UHN (Upper heminephrectomy / partial nephrectomy).

When adopting UU, there are concerns in the literature that include developing reflux, upper pole dysplasia-related hypertension, ureteral strictures, and the need for secondary surgery on the distal ureter.

We hypothesize that lower UU provide a potential benefit over upper heminephrectomy / partial nephrectomy by maintaining the functional upper moiety parenchyma and preventing the kidney pedicle of the duplex system from being dissected, which may result in lower moiety damage, which is in fact to the better viable parenchyma. The aim of this prospective study is to evaluate the short-term effects of lower ipsilateral ureteroureterostomy procedure for the management of ureteric duplication anomalies in children.

Materials and Methods

Following committee ethical approval (IRB: 32769118); a prospective study including patients with complete duplex anomalies presented to our department during the period from March 2023 to March 2024 who had duplex renal system; upper moiety ending with an ectopic ureter or ureterocele and were symptomatic with recurrent febrile UTI, abdominal pain or incontinence and/or images showing affected upper moiety.

All patients in our study underwent thourogh clinical examination, and detailed anatomical radiological examinaiton, including ultrasound renal scan, VCUG,

and MR urography. Functional radiological assesment (DMSA scan) was done as well. Following detailed assessment, parents were consulted about their child condition, the treatment options were offered, and consent for the ureterouretrosotomy and its possible complications was taken.

During surgery, cystosocopy was done at first, to identify the recipient lower pole ureter with the aim of placing a ureteric stent in the lower pole ureter. An inguinal or Pfannenstiel incision was made afterwards. Upper moiety ureter was identified and a distal ureterosotomy is done to create ananstomosis between the upper pole ureter and the lower pole ureter in an end to side fashion.

Postoperative follow up included, renal ultrasound scan after one month and then every three months. In children presenting postoperatively with febrile UTI, assessment with ultrasound and additional MRU is considered depending on the US results for better anatomical details.

Result

Our study was from the period of March 2023 till March 2024 included (32) children with duplex moiety and met the inclusion criteria underwent surgical intervention. A total of twenty-one (21) girls and eleven (11) boys underwent ipsilateral ureteroureterostomy, the age range was from 1 month to 49 months (mean 20.6 months). Urinary tract Infection was the most common presenting symptom in 59.4%, other indications included antenatal hydronephrosis in 28.1% and Incontinence in 12.5 %. **(Table 1 & 2)**

Table 1: The Distribution of the children in the study regarding demographic data (n = 32)

Demographic data	No.	%
Gender		
Male	11	34.4
Female	21	65.6
Age (months)		
Min. – Max.	1.0 –49.0	
Mean ± SD.	20.66 ±12.48	
Median (IQR)	17.0 (12.0 –32.0)	

IQR: Inter quartile range **SD:** Standard deviation

Table 2: The clinical presentation of the children in the study (n = 32)

Presentation	No.	%
UTI	19	59.4
Incontinence	4	12.5
ANH	9	28.1

Regarding the type of pathology; Most of the included cases were ureterocele (75%) and the remaining were ectopic ureter insertion. As regard the follow up of hydronephrosis; After a follow-up of 1 year; hydronephrosis completely resolved in 65.6 % of patients with residual hydronephrosis in 18.8% of patients. **(Table 3)**

Table 3: The Distribution of the children in the study regarding postoperative hydronephrosis (n = 32)

Postoperative hydronephrosis	No.	%
Complete improvement of hydronephrosis	21	65.6
Residual hydronephrosis	6	18.8
Worsening of hydronephrosis	5	15.6

The outcome of UU intervention showed that (84.4%) of patients (7 boys and 20 girls) had successful treatment, while (15.6%) of the children included in the study had adverse events (four boys and one girl).

Median follow up was 10.43 months (IQR:8.1-14.3). Five patients (15.6%) had adverese events postoperatively. Four patients (80.0%) presented with UTI and only one patient had an introperative event of not being able to pass the JJ stent and developed post-operative urinoma, nephrostomy was inserted the second day post-operative then further endoscopic evaluation revealed huge ureterocele ended by open uretrocele excision and reimplanatation since the upper moiety was funcitoing. **(Table 4)**

Table 4: The prevalence of the children in the study who had postoperative adverse events (n = 32)

Early FU	No.	%
No adverse events	27	84.4
With adverse events	5	15.6
Postoperative UTI	4	12.5
Intraoperative events	1	3.1

All these patients pressented initially with ureterocele pathology. In this group, the

four patients who had UTI presented with significant hydropnephrosis on follow up renal scan and needed a seondary surgery. We did a multivariate analysis of the study variables and its relation to the occurrence of adverse events. We found that all patients with adverse events had ureterocele pathology that was not endoscopically incised initally. This was found to be significant (p=0.04). **(Table 5)**

Table 5: The relation between adverse events of the operation and different variables for ureteroureterostomy operation done through one year (2020 –2021) (n = 32)

	Successful treatment (No adverse outcome) (n = 27)		**Adverse treatment outcome (Reoperation, worsening of hydronephrosis or UTI) (n = 5)**		**Test of sig.**	**p**
	No.	**%**	**No.**	**%**		
Gender						
Male	7	25.9	4	80.0	c^2= 5.468*	^{FE}p= 0.037*
Female	20	74.1	1	20.0		
Age (months)						
Min. – Max.	1.0 –49.0		12.0 –33.0		t=0.485	0.640
Mean ± SD.	21.0 ±13.19		18.80 ±8.41			
Median	16.0		18.0			
Presentation						
UTI	15	55.6	4	80.0	c^2=0.821	^{MC}p= 1.000
Incontinence	4	14.8	0	0.0		
ANH	8	29.6	1	20.0		
Ultrasound						
No Hydronephrosis	1	3.7	0	0.0	c^2=0.838	^{MC}p= 1.000
Hydronephrosis of one moiety	24	88.9	5	100.0		

Hydronephrosis of both moiety	2	7.4	0	0.0		
Ureterocele	**19**	**70.4**	**5**	**100.0**	c^2=1.975	^{FE}p= 0.296
Intra vesical ureterocele	14	73.7	1	20.0	c^2= 4.867*	^{FE}p= 0.047*
Ectopic ureterocele	5	26.3	4	80.0		
Ectopic	**8**	**29.6**	**0**	**0.0**	c^2=1.975	^{FE}p=0.296
Ectopic vagina	4	14.8	0	0.0	c^2=0.847	^{FE}p=0.296
Ectopic of upper moiety in UB	1	3.7	0	0.0	c^2=0.191	^{FE}p=1.000
Ectopic in bladder neck	2	7.4	0	0.0	c^2=0.380	^{FE}p=1.000
Ectopic in urethra	1	3.7	0	0.0	c^2=0.191	^{FE}p=1.000
Effect of previous endoscopic evaluation	**(n = 19)**		**(n = 5)**			
No previous endoscopic ureterocele incision	8	42.1	5	100.0	c^2=5.344*	^{FE}p=0.041*
With previous endoscopic ureterocele incision	11	57.9	0	0.0		

Discussion

Options for managing symptomatic or affected duplex renal system include endoscopic procedures, ureteroureterostomy or partial nephrectomy of the nonfunctioning moiety, in addition to the conservative management if indicated.[6] Foley initially reported ipsilateral ureteroureterostomy (UU) as a possible therapy for UD abnormalities in 1928.[16] We believe that UU is a safer procedure than UHN because, in the majority of studies on UU operations, the risk of intraoperative bleeding, blood loss, or damage to the lower moiety is much lower than it is for UHN. In the case of UHN, bleeding may occur if the upper moiety boundaries are not clearly defined and vascular control is not promptly

attained. Vascular injury may also arise when dissecting the upper moiety ureter free of the renal pelvis and hilum.

Also, UU offers a theoretical privilege over UHN as it spares the equivocally functioning upper moiety.[17]

In the past, ureteroureterostomy indications were limited because it was only recommended for children with functional upper moiety to avoid long-term complications like proteinuria, hypertension, and malignant transformation in cases of reserved UM dysplasia. This theory was backed by earlier reports in the literature on the histopathology analysis of samples tested for upper partial nephrectomy. [18-19]

According to a recent publication by Levy et al., who investigated the effects of upper moiety preservation, the pathophysiology of hypertension is primarily closely correlated with renal scarring caused by recurrent febrile urinary tract infections[17] Therefore, it is assumed that the preservation of the UM was not associated with an increased risk of hypertension after a long-term follow-up of approximately fifteen years.[17]

Even in patients with poorly functioning upper moiety parenchyma, upper moiety sparing has been approved in the context of modern conservative follow-up for dysplastic kidneys caused by reflux nephropathy and multicystic dysplastic kidneys.[17]

No significant bleeding occurred during the procedure in our study, according to the literature and the findings of our study on open UU performed on 32 children. Complications from the UU technique are rare and include therapeutic failure, worsening of hydronephrosis, recurring UTIs, and the need to perform secondary surgeries.

Four of the 32 children in our institute who had post-operative adverse events following UU procedures, developed UTIs throughout the follow-up period, according to the results of our study.

We would like to highlight that all patients with adverse events in the postoperative period had initially a ureterocele that was not managed initially. This can raise the question that patients with ectopic ureter benefit more from UU compared to those with ureteroceles especially if not incised initally. Future studies with larger number of patients will be needed to answer this question.

Our results nearly match the results published by Abdelhalim et al,[20] where 27 girls and 8 boys underwent UU, at a median

age of 14.5 months. In this study hydronephrosis improved in approximately ninety percent of operated children. During follow-up of operated children; four patients required further surgical interventions, three had progressive hydronephrosis and seven developed recurrent febrile UTI.

We still consider open UU a challenging procedure that requires a certain level of surgical experiecne with the advantage of minimal blood loss, short operative time, early post-operative recovery with short hospital stay.

At the end of our study we faced some limitation in the form of lacking comparison arm that consequentially limit the ability to compare between the different treatment modalities with sufficient efficacy and power. The number of studied children is relatively low, that is due to the relative scarcity of ureteral duplication requiring surgical intervention, the variety of clinical presentation, other accompanying congenital malformations, the complexity of therapeutic choices, and the absence of firm consensus on the definite decision of management.

Conclusion

Ipsilateral ureteroureterostomy was proved to have low risk of surgical morbidities and low rate of complementary redo surgeries in children with duplex renal anomalies especially those with intravesical ureteroceles and ectopic ureters. More future studies are warranted to assess the long-term outcome of this technique in managing children with duplex renal anomalies as well as comparing it to other surgical options like upper partial nephrectomy.

Ethical Consideration

Approval was obtained from the Ethics Committee of the Faculty of Medicine – Alexandria university, Egypt. (serial Number: 0106601- 19/11/2020)

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Conflict of interests

All authors declare that they have no conflict of interest

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