# Ureteroceles and Ectopic Ureters

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**Primary aims in the diagnosis and treatment of ureteroceles and ectopic ureters:**

1. **To prevent or reduce UTI.**
2. **To prevent renal damage.**
3. **To prevent or correct urinary incontinence.**

## Postnatal Management of Prenatally Detected Ureteroceles and Ectopic Ureters

### Selective Observation of Ureteroceles

**Indications for surgical intervention in ureterocele are febrile UTI (fUTI) and bladder outlet obstruction.**

**One series managing patients by prospective protocol reported that 70 % had resolved or improved hydronephrosis (HN) and VUR, while 23 % had breakthrough UTI and 8 % developed outlet obstruction.**

**Retrospective series with selective observation based on various factors reported that most patients had stable or improved HN and resolved VUR, with <10 % developing breakthrough UTI and <5 % outlet obstruction.**

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A prospective protocol was used to manage 13 patients with ureterocele that was duplex in 11 and single in 2 with MCDK who presented at median age 18 days. Ten were prenatally detected, and three presented with fUTI. Based on either "good" or no function (MCDK) on MAG-3 renography, drainage from the upper pole <30 min, and no bladder outlet obstruction, these patients were all observed with antibiotic prophylaxis (medication not stated). During follow-up at a median of 48 months (25–97), the ureteroceles associated with MCDK were asymptomatic. Upper pole hydronephrosis present in six resolved ($n=3$) or improved to SFU grade 2 ($n=3$), and VUR grade 3 and 4 of the lower pole present in five resolved. The other four had surgery at median 11 months, due to breakthrough UTI in 3 and progressive obstruction in 1 (Han et al. 2005).

Retrospective analysis was done in 52 children with prenatal duplex ureterocele, in which surgical intervention was generally determined by breakthrough UTI, upper pole function >10 %, lower pole obstruction, grade 4 or 5 lower pole reflux, or bladder outlet obstruction. Using these criteria, 14 were observed, eight with VUR, with median follow-up of 8 years (1.6–13). Antibiotic prophylaxis (medication not stated) was used "routinely until toilet training" or age 5, if there was VUR. No patient developed UTI, VUR resolved in 3/4 cases with cystography, and hydronephrosis was stable ($n=8$) or improved/resolved with collapse of the ureterocele ($n=6$) (Shankar et al. 2001).

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A retrospective study found 10 newborns, six females, with prenatally detected ureteroceles who had "adequate drainage of the affected unit" without lower pole obstruction, grade 4 or 5 VUR, or bladder outlet obstruction managed with antibiotic prophylaxis (medication not stated) and observation. Six were duplex and 4 single-system ureteroceles, 1 with MCDK. During mean follow-up of 3 years, HN (initially SFU grade 3 in 2 and less in the others) resolved in six and was improved or stable in 4. VUR grade 3 into the lower pole was initially present in 4, which resolved in 2. No patient underwent repair (Direnna and Leonard 2006).

Another retrospective analysis included 40 neonates with prenatally detected ureterocele initially observed with antibiotic prophyaxis (medication not stated) with planned open surgery at 6 months (selection for observation vs. puncture was not stated, but implied these had no lower pole VUR and no bladder outlet obstruction). During observation, 3 (8 %) had UTI and 2 (5 %) had progressive ureterocele enlargement resulting in bladder outlet obstruction (Husmann et al. 2002).

## UTI in Newborns

**No trials compare antibiotic prophylaxis versus no treatment for newborns with prenatally diagnosed anomalies, including ureteroceles or ectopic ureters.**

**One small series of neonates with prenatal ureterocele reported that 53 % had UTI with no difference in those with versus without antibiotic prophylaxis.**

**A study in which newborns with ureteroceles with VUR were punctured and those without VUR observed found breakthrough UTI in 8 % of both groups by age 6 months.**

**Another series that used antibiotic prophylaxis in newborns with ureterocele or ectopic ureter without VUR reported that 42 % developed fUTI at median 3 months of age.**

No trial has compared antibiotic prophylaxis versus no treatment in neonates with prenatally detected ureterocele or ectopic ureter.

Retrospective review concerned 15 neonates, 13 females, with prenatally detected ureterocele that was duplex in 14 cases and associated with VUR in 10. Of these, 10 were evaluated within the first week of life, 3 in the second, and 1 at 3 and 6 weeks each. FUTI occurred in seven (47 %) due to *E. coli* ($n=6$) and *S. aureus*, in three despite prophylactic antibiotics (medication not stated). There was no difference in UTI in those with versus without antibiotic (43 % vs. 62.5 %, $p=0.6$) (Besson et al. 2000).

Another retrospective study involved 72 neonates with prenatally detected duplex system ureteroceles all treated from birth with antibiotic prophylaxis (medication not stated). Of these, 32 had endoscopic puncture at a median age of 5 days, while the other 40 were initially observed but had open surgery at ≤6 months of age (median 3 months, 2–6). UTI occurred after puncture and before age 6 months in 3/32 (9 %) in 1 with bladder outlet obstruction from incomplete decompression, and in 3/40 (8 %) with delayed intervention (Husmann et al. 2002).

The protocol used by Han et al. (2005) described above reported that 3/11 (27 %) neonates with duplex ureteroceles selected for nonsurgical management in part because they were considered to be at low risk for infection had breakthrough UTI, despite antibiotic prophylaxis at median time 11 months.

The ten patients observed by Direnna and Leonard (2006) described above, with 6 duplex and 4 single-system ureteroceles, reported no breakthrough UTI during mean follow-up of 3 years. Antibiotic prophylaxis was used in all for a mean of 1.5 years.

Retrospective review of consecutive neonates treated with a systematic protocol included 12 prenatally detected with duplex ectopic ureter or ureterocele without VUR and initially treated with antibiotic prophylaxis (medication not stated). Five (42 %) developed breakthrough fUTI at median age 3 months (2–8), three with ectopic ureter and 2 with ureterocele (Prieto et al. 2009).

## Endoscopic Management of Ureterocele

**Secondary surgery after ureterocele puncture is reported more likely with extravesical ureteroceles, duplex systems, and pre-puncture VUR.**

**One retrospective comparison of puncture versus upper pole heminephrectomy for duplex ureterocele with no VUR reported secondary surgery more likely after puncture, mostly for new VUR.**

**Indications for secondary surgery often are not reported.**

Systematic literature review was done to determine secondary operation rates after ureterocele puncture, reporting additional surgery was more likely with:

- Extravesical ureterocele, RR 2.8 (95 % CI 2.06–3.74).
- Duplex system, RR 3.9 (95 % CI 1.14–10.93).
- Pre-puncture VUR, RR 1.56 (95 % CI 1.24–1.96).

The authors used secondary surgery as the endpoint, but indications for both initial puncture and subsequent surgery were not always reported in reviewed articles (Byun and Merguerian 2006).

Retrospective review was done in 52 children, 42 % female, undergoing ureterocele puncture done at median age 3 months (1 month–12 years). This series dated from 1984 to 2001, and only 12 (23 %) had prenatal diagnosis, while the others presented after UTI. Ninety-two percent had ureteral duplication, and the 52 cases were classified as intravesical in 73 % and extravesical in 27 %. Antibiotic prophylaxis (not described) was used for all with VUR until resolution. Preoperative and postoperative nuclear renography was done (postoperative time interval not stated). Nonfunctioning upper poles ($n=10$) or kidneys ($n=9$) were removed. Median follow-up was 9 years (6 months–18 years):

- Poor function in the upper pole ($n=25$) did not improve, and normal to moderately impaired function did not decrease after puncture and during follow-up ("moderate" and "poor" were not defined; actual data not shown).
- No patient with poor function had pyelonephritis.
- VUR spontaneously resolved in 59 % of affected renal units; others mostly had endoscopic correction. Indication for VUR treatment was not stated.
- No patient had incontinence.
- No patient had infection after puncture (Chertin et al. 2003).

Another retrospective series included 60 children, 68 % female, with ureteroceles treated with puncture: duplex in 51 that was intravesical in 22 and extravesical in 29, and single system in nine with intravesical ureterocele. Thirty-two (52 %) presented before 3 months of age, and 31 (52 %) were prenatally detected, while 29 (48 %) had fUTI. With follow-up a mean of 20 months (4–62), 19 (32 %) had additional surgery.

- None of the nine single-system ureteroceles had additional surgery, and of four of nine with new VUR after puncture, three resolved.
- Of 51 duplex ureteroceles, 19 had secondary surgery, including seven (14 %) with "persistent VUR and infection."

Otherwise, indications for secondary surgery were not described (Hagg et al. 2000).

A third retrospective series of 28 patients with duplex ureterocele and no VUR that underwent puncture reported new VUR in 16 (57 %) and persistent obstruction in 2 (7 %). During follow-up a median of 2 years (3 months–6 years), reimplantation was done in 12 for breakthrough UTI or persistent reflux. Compared to partial nephrectomy in patients with similar preoperative findings (duplex ectopic ureterocele and no VUR), puncture was significantly more likely to result in further surgery or persisting VUR, 64 % versus 15 % (see below) (Husmann et al. 1999).

## Upper Tract Surgery for Ureterocele and Ectopic Ureter

**Retrospective series report secondary surgery in <20 % for VUR or symptomatic remnant upper pole stumps (see below).**

In a retrospective review, 26 patients with duplex ureterocele and no VUR had partial nephrectomy as initial therapy. New lower pole VUR occurred in eight (30 %), which then resolved in 4 and led to reimplantation in 4 (15 %) due to breakthrough UTI during median follow-up of 2 years (1–4). These outcomes were compared to puncture in similar patients, and it was concluded that heminephrectomy results in fewer secondary operations (see section above) (Husmann et al. 1999).

Another review included 29 patients with 30 ureteroceles, six with VUR, who underwent

upper pole heminephrectomy ($n=29$) or pyelopyelostomy and then had follow-up for a mean of 25 months (9–30). New VUR occurred in 4 and then resolved in 3, and 4 (14 %) others developed breakthrough UTI leading to lower tract surgery (Gomes et al. 2002).

An upper tract approach was used in 31 duplex systems with ectopic ureters in 30 females and one male as newborns to age 17 years. Of these, 23 systems were considered on various imaging studies (IVP, CT, nuclear renography) to have poor or no function and had heminephrectomy, while the other eight had "sufficient function to warrant salvage" (not defined) and so had high ureteroureterstomy or ureteroplyelostomy. The orifice of the ectopic ureter was identified in 19 (58 %), at the bladder neck or urethra in 15, and vagina or vestibule in 4. During follow-up a mean of 4.5 years (8 months–10 years), 3 (10 %) had fUTI leading to remnant stump removal (subsequent clinical course not described), and 1 had pain with voiding, resulting in stump removal (see below). Preoperative VUR occurred in nine (29 %), into the ipsilateral lower pole in 5 and contralateral in 4. One of these had reimplantation for "persistent reflux," while outcomes of VUR in the others were not stated (Plaire et al. 1997).

Laparoscopic heminephrectomy was done in 17 consecutive patients with 19 affected units having ectopic ureter ($n=8$) or ureterocele ($n=7$), of which three had lower pole VUR. Follow-up was a mean of 57 months (8–115), during which time lower pole VUR resolved in 3/4 units. The only infections involved the stump and occurred in three patients (18 %) leading to removal. One patient had loss of function to the remaining lower pole. Two of seven with ureteroceles had persistent, but smaller, ureteroceles that were asymptomatic (Denes et al. 2007).

## Lower Tract Surgery for Ureterocele

**Ureterocele excision or unroofing with reimplantation, sometimes following prior puncture, resulted in new or persistent VUR in 12 %, reported by two series.**

**Postoperative fUTI was reported in 0–10 %.**

**Complications from retained upper pole segments did not occur.**

Fifty-seven patients, 94 % female, at mean age approximately 28 months, underwent ureterocele surgery at three institutions by four surgeons based on their preference. Thirty-nine (68 %) had prior puncture, and operations comprised ureterocele excision or unroofing and ureteral reimplantation. During mean follow-up of 55 months (6–234), new VUR occurred in seven (12 %) and fUTI in six (11 %), of which two had reflux. Outcomes for the new VUR were not described (Lewis et al. 2008).

A review was done in 16 children, 15 females, with duplex ureteroceles and nonfunctioning upper poles who had lower tract reconstruction without upper pole excision at two institutions. Thirteen had prior puncture, and all had ureterocele excision, bladder neck reconstruction, and reimplantation. Follow-up was a mean of 62 months (33–127). VUR was found in 2 (12.5 %), 1 observed and the other not further described, and there were no fUTIs. The retained nonfunctioning upper pole moiety did not cause hypertension or other recognized complication (Gran et al. 2005).

Another review included 31 children, 28 females, mean age 30 months (19 days–10 years) with duplex ectopic ureters. Twelve with functioning upper poles (visualization in IVP) had ureteral reimplantation of the ectopic ureter, while 18 with non-visualization had upper pole heminephrectomy; one kidney had no function. During follow-up a mean of 66 months (6 months–20 years), one patient in each treatment group had recurrent UTI (not otherwise described). None had incontinence, and none with reimplantation had further surgery (El Ghoneimi et al. 1996).

## Simultaneous Upper and Lower Tract Reconstruction for Ureterocele

**One study reported secondary surgery after "complete" reconstruction in 6 % of patients.**

Retrospective review was done in 18 patients with 20 ureteroceles who all had VUR and underwent upper heminephrectomy ($n=11$) or upper pole ureteral tailoring ($n=9$) and ureterocele excision, bladder neck reconstruction, and cross-trigonal reimplantation. Duration of follow-up was not stated (mean 33 months for all patients in the series); new contralateral VUR occurred in 1, and two patients had postoperative complications, including bilateral obstruction after bilateral reimplantation and one requiring intermittent catheterization. There was no mention of postoperative UTI; 1 (6 %) patient had reoperation (not further described) (Gomes et al. 2002).

## Ureteroureterostomy

**Two series using different inclusion criteria reported secondary surgery in <10 % of children after ureteroureterstomy (UU).**

Of 142 children undergoing surgery for duplex ureters, 39 (17 %), 86 % females, underwent 41 UUs. There were five with ectopic ureter and four with ureterocele who only had UU. Nine others with ectopic ureters with lower pole reflux had UU and reimplantation, as did another 12 with prior ureterocele puncture. The remaining patients had lower pole reflux without ectopic ureter or ureterocele. Mean follow-up was 12 months (3–34). Two cases of new contralateral VUR were corrected with endoscopic injection, as was one of two cases with ipsilateral persistent VUR, comprising secondary interventions in 8 %. There was no mention of postoperative UTI and "all showed improvement of dilation" on ultrasound (Chacko et al. 2007).

Another review included 23 consecutive children, (74 %) female, and 26 duplex systems with ectopic ureter ($n=18$) or ureterocele ($n=8$) and no lower pole reflux, who represented 37 % of patients with these conditions operated during a 4-year study period. Median patient age was 10 months (2–56), and two of seven patients with ureteroceles had prior puncture with resultant upper pole reflux. Mean follow-up was 26 months (2–48). Preoperative upper pole hydronephrosis occurred in 22 ureters and was SFU grades 3 and 4 in 13 (50 %); postoperative ultrasound at 12 weeks showed resolution ($n=17$) or reduction to grade 2 or less in all ureters. One patient had fUTI and was found to have new grade 1 ipsilateral VUR treated with endoscopic injection, for secondary surgery rate of 4 % (Prieto et al. 2009).

## Renal Function

**Our review found only two studies mentioning renal functional outcomes. One stated that 8 % of patients had a decrease >10 % in renographic differential renal function after heminephrectomy.**

From a retrospective review of 101 patients with heminephrectomy renal function, data were available in 60, of which 42 had upper pole (ureterocele in 20) and 18 lower pole excision. Mean age at operation was 44 months, and follow-up was a mean of 25 months (3–64). Preoperative and postoperative MAG-3 or DMSA scans were obtained (date of postoperative study not stated). Mean preoperative differential function was 40 % (22–61 %), and 33 % (13–60 %) postoperatively (statistical analysis not reported). Five (8 %) had a decrease >10 %—one who had recurrent postoperative UTI and without known reason in others (Gundeti et al. 2005).

The retrospective study by Gran et al. (2005) described above, in which bladder but not renal level surgery was done, reported that no patient had loss of renal function by nuclear renography, but timing of studies and resultant data were not stated or shown.

## Histology of Nonfunctioning Renal Segments

**Histology of resected upper pole segments was reported in two studies to most often demonstrate inflammatory changes or dysplasia.**

Retrospective review of the upper pole was done in 50 consecutive heminephrectomy specimens related to ureterocele ($n=30$) or ectopic ureter. Twenty-five demonstrated nonspecific inflammatory findings in the interstitial tissues,

chronic inflammatory changes with collecting system involvement in 15, microabscesses in 3, xanthogranulomatous pyelonephritis in 2, and dysplasia (parenchymal disorganization with cysts, primitive ducts, cartilage) in 30 with nephroblastomatosis in 1. Only one was interpreted as normal. There were no apparent differences in patients with antenatal versus postnatal diagnosis, but age at surgery and presurgical morbidities, including fUTI, were not described. The implication of these findings, however, was that there was little expectation that earlier diagnosis and treatment would significantly improve upper pole findings (Abel et al. 1997).

The 16 upper pole specimens with ectopic ureters removed for non-visualization on IVP by El Ghoneimi et al. (1996) as described above demonstrated "lesions of chronic pyelonephritis" in 13 and "focal lesions of dysplasia" in 4.

The practice of routinely removing upper poles based on such histologic findings was challenged by a review article that addressed association of renal dysplasia to UTI, hypertension, and tumor potential and concluded that occurrence of these is so rare that it does not support extirpation (Husmann 1998).

## Continence

**One study reported no postoperative incontinence after surgery for ectopic ureter.**

Of the 28 females with duplex ectopic ureters reported by El Ghoneimi et al. (1996) above, the upper pole ureter entered the bladder neck in six, posterior urethra in 5, vagina in seven, vestibule in six, and was unclear in 4. Preoperative incontinence occurred in 13 (46 %), corrected in all cases by either upper pole reimplantation or heminephrectomy.

Of 31 duplex systems with ectopic ureters in 30 females and one male reported by Plaire et al. (Plaire et al. 1997) above, the orifice of the ectopic ureter was identified in 19 (58 %), at the bladder neck or urethra in 15, and vagina or vestibule in 4. The authors did not mention postoperative incontinence.

## Bladder Dysfunction

**There are no reliable data to determine if children with ureteroceles and ectopic ureters have increased bladder dysfunction compared to normal children.**

A retrospective series analyzed 34 children, 79 % females, after ureterocele management. Eighteen (53 %) had extravesical ureteroceles. Median age at surgery was 10 months (3–101) and involved heminephrectomy for ten, ureterocele excision and reimplantation in six, both upper and lower tract surgery in 15, and no incision or no treatment in 3. Of these, 32 had bladder function assessment at median 5 years of age (1–12). Infrequent voiding <4×/daily was diagnosed in 19 (59 %). Another three (9 %) had incontinence, none with prior bladder surgery, including one with a persistent ureterocele. Two (6 %) used CIC because of "high" PVR. UD was done in 27 children, with 55 % having measured capacity >150 % predicted for age and 11 % detrusor instability; all had normal compliance and mean PVR was 12 cc. Uroflow pattern was stated to be normal in all cases. The authors considered only eight (25 %) to have normal bladder function defined as normal capacity and PVR <5 cc, but they did not correlate factors they considered abnormal to clinical symptoms or complications, such as UTI (Abrahamsson et al. 1998).

The four-surgeon series with ureterocele excision and reimplantation by Lewis et al. (2008) described above reported voiding dysfunction (infrequent voiding, holding maneuvers, urgency) postoperatively in 11 (20 %). One other child had incontinence, possibly due to bladder neck dysfunction.

## Symptomatic Stumps

**Three retrospective series report removal of the remnant "stump" after partial upper pole ureterectomy in from 7 to 18 %, mostly due to UTI.**

**None reported postoperative UTI occurrence after stump excision.**

Stump removal was done in 4 (7 %) of 55 patients with duplex ureters with ureterocele, ectopia, and/or VUR who underwent upper pole heminephrectomy as described by Ade-Ajayi et al. (2001) above. All had recurrent UTI (not described), but follow-up after removal and UTI occurrence, if any, in other patients with stumps was not stated.

Of the 32 patients described by Plaire et al. (1997) with duplex ectopic ureters who all had upper tract surgery (heminephrectomy in 23 and reconstruction in eight), 4 (12 %) had secondary surgery to remove the remnant stump, 1 during a reimplantation for persistent lower pole reflux, 1 for pain during voiding, and two for fUTI.

Laparoscopic heminephrectomy was done in 17 consecutive patients with 19 affected units having ectopic ureter ($n=8$) or ureterocele ($n=7$). Follow-up was a mean of 57 months (8–115), during which time infections involved the stump in three patients (18 %) and were removed (Denes et al. 2007).

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