

# Management of duplex system ureteroceles in neonates and infants

Marco Castagnetti and Alaa El-Ghoneimi

**Abstract** | Ureteroceles associated with the upper pole of a complete ureteral duplication are known as duplex system ureteroceles (DSUs). A limited knowledge of the natural history of this condition makes its management in neonates and infants controversial. Asymptomatic neonates diagnosed antenatally in the absence of severe hydronephrosis (HUN) are at low risk of developing urinary tract infections during the first months of life. These patients might, therefore, begin antibiotic prophylaxis and undergo comprehensive assessment by 3–6 months. Subsequently, conservative management can be viable in patients without severe HUN or high-grade vesicoureteral reflux (VUR). Cases of DSUs with severe HUN but no VUR can be treated by an upper urinary tract approach or by endoscopic decompression depending on upper pole function. Patients with preoperative VUR represent the most challenging cases. If VUR cure is considered necessary, lower urinary tract reconstruction is recommended. Endoscopic decompression allows for a definitive treatment in at least 50% of cases and, in the remaining cases, can be combined with conservative management or endoscopic treatment of VUR. Nonfunctioning or poorly functioning upper poles can be left *in situ* both in lower urinary tract reconstruction and after endoscopic decompression. Parental preferences should also be taken into account in the decision-making process.

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### Learning objectives

Upon completion of this activity, participants should be able to:

- 1 Identify the prevalence of urinary tract infection in duplex-system ureteroceles (DSUs) and the means to prevent infection among these patients.
- 2 Specify the complications of DSUs.
- 3 Describe the techniques and efficacy of endoscopic decompression of ureteroceles.
- 4 Identify the recommended treatment of a child with DSUs and severe hydronephrosis.

### Competing interests

The authors, the Journal Editor A. Hay and the CME questions author C. P. Vega declare no competing interests.

## Introduction

The widespread use of ultrasonography has changed the diagnostic pattern of ureteroceles (Figure 1) such that up to 75% of cases are currently diagnosed antenatally or in early infancy;<sup>1–4</sup> in the vast majority of cases, the ureterocele affects the upper moiety of a duplex system.<sup>1,2,5</sup> Initial management of duplex system ureteroceles (DSUs) in neonates and infants should aim to decrease the risk of urinary tract infections (UTIs), preserve the function of the lower moiety and contralateral kidney, and avoid bladder dysfunction and incontinence.<sup>6</sup>

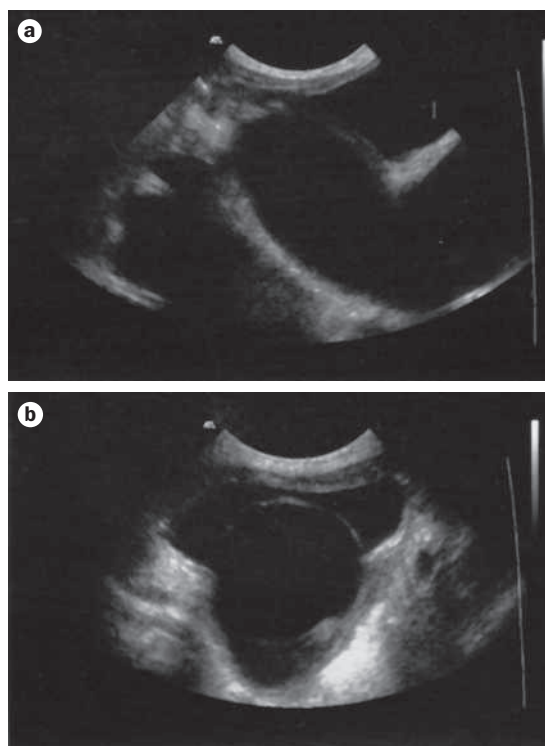
To fulfill these goals, different approaches have been proposed, including total reconstruction (upper pole heminephrectomy, ureterocele excision, bladder base/neck reconstruction, and lower-pole ureter reimplantation), the upper tract approach (ureterocele moiety heminephrectomy or pyelopyelostomy), and endoscopic ureterocele decompression. Endoscopic decompression has become popular as a minimally invasive means of achieving timely ureterocele decompression and preventing UTIs, while avoiding extensive trigonal surgery, which risks damaging bladder function in infants;<sup>1,4–7</sup> by contrast, total reconstruction has been recommended as the most effective approach to achieve single-stage cure of DSUs and prevent long-term bladder dysfunction and incontinence.<sup>2,8</sup> Flexible strategies have also been proposed, with approaches chosen depending on preoperative variables, such as ureterocele position (intravesical versus ectopic), and number of moieties with vesicoureteral reflux (VUR) or non-refluxing hydronephrosis (HUN).<sup>9–11</sup> Finally, conservative

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**Key points**

- The risk of urinary tract infections in patients receiving antibiotic prophylaxis is <10% before 6 months of age; asymptomatic neonates and infants without massive upper tract dilatation can be treated conservatively
- Ureterocele moiety function is generally negligible; scintigraphic assessment is reliable; nonfunctioning poles can be left *in situ*
- Preoperative vesicoureteral reflux tends to persist after ureterocele decompression, but <50% of patients with reflux develop urinary tract infections
- Incontinence is rare in patients with ureteroceles; it is unrelated to initial treatment modality, and seems to be refractory to both medical therapy and bladder-neck reconstruction
- Endoscopic decompression is the approach of choice for intravesical ureteroceles, and is the definitive treatment in 50% of ectopic ureteroceles; it can also be combined with conservative or endoscopic management of persistent reflux
- Total reconstruction can be performed safely (also in infancy); the need for upper pole partial nephrectomy and systematic reconstruction of the bladder base and neck is disputed



**Figure 1** | Neonatal ultrasound scan. **a** | Severe upper urinary tract dilatation. **b** | Ureterocele presence within the bladder.

management has been proposed for selected cases of DSUs.<sup>12–16</sup>

In the absence of controlled trials, however, the choice of initial management can only be based on the balance between the potential risks inherent to the condition and the results of each treatment modality. Here, we aim to assess these two aspects and, hence, discuss possible strategies for the initial management of DSUs in neonates and infants.

**Aspects inherent to the condition**

**UTIs before treatment**

When a patient presents with urosepsis, timely endoscopic decompression is generally the consensus approach.<sup>17</sup> However, the widespread use of ultrasonography and appropriate antibiotic treatment has made cases of urosepsis increasingly rare, and the major clinical issue is, rather, whether early decompression prevents UTIs in asymptomatic neonates or recurrent infections in symptomatic cases. Prevention of infections is key to preserving renal function and preventing long-term untoward outcomes, such as hypertension.<sup>18</sup>

Data regarding infection rates in asymptomatic neonates with ureteroceles diagnosed antenatally are inconsistent; whereas Besson *et al.*<sup>19</sup> reported an infection rate of 50%, Decter *et al.*<sup>2</sup> failed to observe UTIs in any of their patients awaiting surgery. Infection rates have been reported to increase with age, from 3% in patients younger than 3 months<sup>20</sup> to 12% by 6 months of age.<sup>9</sup> Husmann *et al.*<sup>17</sup> found comparable infection rates after 6 months' follow-up of 32 neonates who had undergone early endoscopic decompression and 40 neonates who were treated with antibiotic prophylaxis and delayed surgery (9% and 8%, respectively). Patients in this study were, however, allocated to each group based on physician preference, which potentially causes a selection bias, with decompression favored for more severe cases.

Thus, although the number of patients studied overall is small, the evidence seems to refute the hypothesis that systematic ureterocele decompression is necessary in order to prevent UTIs, at least in asymptomatic neonates and infants without high-grade HUN or VUR. The latter, instead, are generally considered to mandate primary surgical management.<sup>13,15,16</sup>

Antibiotic prophylaxis is generally recommended during conservative management,<sup>21</sup> although published data are, again, quite variable. Direnna and Leonard<sup>15</sup> administered prophylaxis for a mean of 1.5 years; Coplen and Austin,<sup>13</sup> until resolution of VUR or completion of toilet training; and Shankar and colleagues,<sup>12</sup> until the age of 5 years in cases of persistent VUR.

**Ureterocele moiety function**

Preservation of the function of the upper pole has classically been considered a factor supporting early ureterocele decompression, as the obstruction was deemed to impair reliable scintigraphic assessment and decompression was thought to facilitate recovery of function.<sup>6,22</sup> The ability to salvage ureterocele moiety function has been investigated by renal scintigraphy<sup>23–25</sup> or by histologic analysis of the removed pole after heminephrectomy.<sup>26–28</sup>

Nonfunctioning or poorly functioning upper poles have been reported in 60–100% of patients with DSUs.<sup>3,4,18,29</sup> Connolly *et al.*<sup>23</sup> found an average upper pole differential function of only 3% in 20 neonates with DSUs; Vates *et al.*<sup>24</sup> reported a mean decrease in function of 1.25% after heminephrectomy and Gundeti *et al.*<sup>25</sup> noted a substantially unchanged function in 19 of 20

**Table 1** | Rates of preoperative VUR in non-ureterocele-bearing moieties in duplex system ureteroceles

Study	Patients (n)	Cases of preoperative VUR (n)	Intravesical ureteroceles (n)	Ectopic ureteroceles (n)
Petit <i>et al.</i> (1999) <sup>29</sup>	18	8 (44%)	1 (12.5%)	7 (87.5%)
Castagnetti <i>et al.</i> (2004) <sup>7</sup>	41	20 (49%)	7 (35%)	13 (65%)
Hagg <i>et al.</i> (2000) <sup>1</sup>	51	27 (53%)	ND	ND
Upadhayay <i>et al.</i> (2002) <sup>3</sup>	89	52 (58%)	21 (40%)	31 (60%)
Husmann <i>et al.</i> (1999) <sup>42</sup>	165 <sup>a</sup>	111 (67%)	NA	NA
Jayanthi and Koff (1999) <sup>22</sup>	21 <sup>a</sup>	14 (67%)	NA	NA
Chertin <i>et al.</i> (2005) <sup>4</sup>	48	35 (73%)	ND	ND
Decter <i>et al.</i> (2001) <sup>2</sup>	36 <sup>a</sup>	27 (78%)	NA	NA

<sup>a</sup>All duplex system ureteroceles were ectopic. Abbreviations: NA not applicable; ND, not determined; VUR, vesicoureteral reflux.

**Table 2** | Rates of persistent VUR after decompression of duplex system ureteroceles

Study	Approach	Preoperative VUR	Postoperative VUR	Persistent VUR
<b>VUR persistence by patients</b>				
Upadhayay <i>et al.</i> (2002) <sup>3</sup>	UPPN <sup>a</sup>	21	13	62%
Husmann <i>et al.</i> (1999) <sup>42</sup>	UPPN	67	56	84%
Husmann <i>et al.</i> (1999) <sup>42</sup>	ED	44	37	84%
Petit <i>et al.</i> (1999) <sup>29</sup>	ED	7	6	86%
Jayanthi and Koff (1999) <sup>22</sup>	ED	14	13	93%
<b>VUR persistence by RUU</b>				
Chertin <i>et al.</i> (2005) <sup>4</sup>	ED	44	27	61%
Castagnetti <i>et al.</i> (2004) <sup>7</sup>	ED	20	15	75%
Kajbafzadeh <i>et al.</i> (2007) <sup>44</sup>	ED	29	33	113%

<sup>a</sup>Patients undergoing endoscopic treatment were not included because postoperative reflux rate also included *de novo* reflux in the ureterocele moiety. Abbreviations: ED, endoscopic decompression; RUU, renoureteric units; UPPN, upper pole partial nephrectomy; VUR, vesicoureteral reflux.

patients who underwent upper pole heminephrectomies. Notably, rates of nonfunctioning or poorly functioning upper poles were comparable between prenatally and postnatally diagnosed cases,<sup>3,4</sup> which suggests that functional impairment is not progressive. Impairment of function also seems to be irreversible. Connolly *et al.*<sup>23</sup> repeated their scintigraphic analysis in nine of their study cases after endoscopic decompression and observed no functional improvement. Vates *et al.*<sup>24</sup> observed a mean increase in differential function of 2.25% after upper pole salvage (by pyelopyelostomy or ureteroureterostomy).

Nevertheless, upper pole function can sometimes make a significant contribution. In the series by Levy and colleagues,<sup>18</sup> 25% of 115 cases of DSUs had a differential function of >15%, whereas 4 of the 20 patients assessed by Connolly *et al.*<sup>23</sup> had a differential function of 7–19%.

With respect to histology, around 70% of removed upper poles show evidence of dysplastic changes. Again, there is no obvious difference in this figure between antenatally and postnatally diagnosed cases,<sup>26–28</sup> which further indicates that renal damage in patients with DSUs is primarily congenital and cannot improve over time or after ureterocele decompression.

The ultimate issue in clinical practice is whether preoperative assessment might reliably predict the degree of renal damage. Levy *et al.*<sup>18</sup> reported renal dysplasia and/or chronic pyelonephritis in 89% of patients undergoing heminephrectomy for a nonfunctioning ureterocele moiety. Bolduc *et al.*<sup>30</sup> subjected many features of preoperative radiological assessment to multivariate analysis, and found severe parenchymal thinning on ultrasonography and minimal function on preoperative scintigraphy to be accurate predictors of severe histologic lesions.

Finally, it is noteworthy that nonfunctioning upper moieties left *in situ* have seldom been reported to cause symptoms or increase long-term morbidity of the condition, irrespective of the chosen treatment approach.<sup>4,31,32</sup>

#### Preservation of associated renal units

The ureterocele can put the lower pole or contralateral units at risk of damage by obstructing either unit or the bladder outlet. These conditions are both uncommon, but there is general agreement that the presence of obstruction, detected as non-refluxing or progressive dilation of associated moieties or as an alteration in renal function, mandates timely ureterocele decompression.<sup>17</sup>

**Table 3** | Rates of postoperative urinary tract infections

Study	Approach	Mean/median follow-up (years)	Patients (n)	Postoperative UTIs (n)
Chertin <i>et al.</i> (2005) <sup>4</sup>	ED	9	48	3 (6%)
Shekarriz <i>et al.</i> (1999) <sup>9</sup>	ED/UPPN/total reconstruction	5–7	72	6 (8%)
Beganovic <i>et al.</i> (1999) <sup>37</sup>	Total reconstruction	9.6	55	9 (17%)
Upadhyay <i>et al.</i> (2002) <sup>4</sup>	ED/UPPN/BS	3.5	95	21 (22%)
Chertin <i>et al.</i> (2001) <sup>33</sup>	ED	6	34	8 (23.5%)
Singh and Smith (2001) <sup>34</sup>	ED	ND	24	6 (25%)
Chertin <i>et al.</i> (2007) <sup>38</sup>	ED	4.2	46	12 (26%)
Sauvage <i>et al.</i> (2003) <sup>35</sup>	ED/UPPN/total reconstruction	6	126	33 (26%)
Husmann <i>et al.</i> (1995) <sup>20</sup>	UPPN	2	87	28 (32%)
Lewis <i>et al.</i> (2008) <sup>36</sup>	Lower urinary tract reconstruction	4	54	24 (44%)
Jayanthi and Koff (1999) <sup>22</sup>	ED	3	21	10 (47%)

Abbreviations: BS, bladder surgery; ED, endoscopic decompression; ND, not determined; UPPN, upper pole partial nephrectomy.

VUR is usually considered the major risk factor for ipsilateral or contralateral moieties. VUR occurs in 50–75% of patients with DSUs, and is more commonly associated with ectopic than intravesical ureterocele (Table 1). VUR seldom ceases after ureterocele decompression (Table 2), particularly when it is into the ipsilateral lower moiety, perhaps because it reflects an irreversible anatomical distortion of the trigonal anatomy. VUR is considered to put ipsilateral and contralateral moieties at risk of damage because it is a risk factor for UTIs. Any definite correlation between VUR and UTIs, however, remains largely unknown; accurate data do not exist because febrile and nonfebrile, and isolated and recurrent UTIs are all grouped together in many series. Our review, however, suggests that UTIs occur after treatment in about 25% of patients with DSU (Table 3)—more commonly in postnatally diagnosed cases.<sup>3,4</sup> UTIs occur in about 50% of patients with postoperative VUR,<sup>20,33</sup> but can also occur in the absence of VUR.<sup>32,34–37</sup> Consistently, UTIs have also been reported after lower urinary tract reconstruction.<sup>8,9,37</sup>

The risk of developing a UTI might not be equal in all patients with ureterocele. Upadhyay *et al.*<sup>3</sup> reported that, after one UTI, patients might be intrinsically more prone to develop recurrent bouts, irrespective of the surgical management. According to Chertin *et al.*,<sup>38</sup> instead, patients with low-grade VUR or VUR downgraded following endoscopic decompression might be less likely to develop UTIs and could be withdrawn from prophylaxis.

#### Bladder dysfunction and continence

Bladder dysfunction, caused by defective detrusor activity, and urinary stress incontinence, as a result of a muscular defect at the bladder neck, can occur in patients with DSUs. Both these outcomes have been reported only

in patients with ectopic ureterocele,<sup>39–41</sup> and can occur after any kind of treatment, which suggests a congenital, rather than iatrogenic, origin.<sup>35,39,40</sup>

In fact, there is no consensus on the frequency and type of bladder dysfunction that is associated with ureterocele. Some authors have reported that the majority of patients present with high-capacity bladders and suffer from incomplete emptying, leading to the postulation that ureterocele primarily cause a failure of myogenic detrusor muscles.<sup>39,41</sup> Other authors, instead, reported lower urinary tract symptoms and incontinence in only a minority of cases, with urgency as the most frequent symptom.<sup>40</sup> Irrespective of the type of bladder dysfunction, about 90% of affected patients were reported to respond promptly to behavioral modifications and medical therapy.<sup>40</sup>

Stress urinary incontinence, instead, seems to be quite a rare occurrence in ureterocele patients, but can be quite troublesome to treat when it occurs.<sup>20,39</sup> Indeed, it seems to be unresponsive to medical treatment and bladder-neck reconstruction. In the series by Husmann *et al.*,<sup>20</sup> two of four patients undergoing bladder-neck reconstruction required the eventual placement of an artificial urinary sphincter; in another series, of four incontinent patients, two required an artificial urinary sphincter and the other two required bladder-neck closure.<sup>40</sup> In contrast, Merguerian *et al.*<sup>16</sup> reported normal voiding and complete bladder emptying in 12 toilet-trained children who underwent lower urinary tract surgery without ureterocele excision. To our knowledge, the best study currently available that addresses the possible correlation between the trigonal defect owing to the ureterocele and the risk of incontinence is a case-control study comparing patients who underwent ureterocele excision or marsupialization; again, no difference was observed between patients treated by either approach.<sup>36</sup>

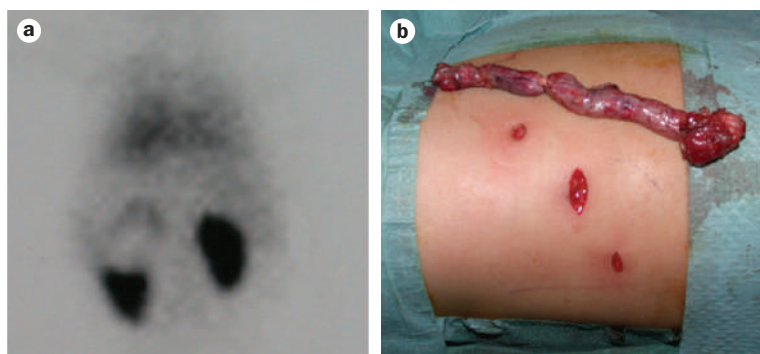
## Results of management options

### Endoscopic decompression

Endoscopic management is the easiest way to achieve ureterocele decompression in neonates and infants. Many series have assessed the effectiveness of decompression in relation to ureterocele position: it seems to be the approach of choice for intravesical DSUs,<sup>5,42</sup> whereas it comprises the definitive treatment for about half of ectopic ureteroceles.<sup>3,7</sup> Technical reasons might partly explain the different success rates.

According to current consensus, the best technique involves making an incision or puncture in the intravesical portion of the ureterocele, just above the level of the bladder wall,<sup>1,5,7,33,34,42</sup> to create a valve mechanism that prevents the development of VUR in the punctured moiety. Opening of the extravascular portion of the ureterocele is no longer recommended, as it only increases the risk of *de novo* reflux in the punctured moiety.<sup>1,6</sup> Several instruments can be used for the procedure: bugbee electrodes, cold knives, metal catheter guides, and lasers.<sup>43</sup> Kajbafzadeh *et al.*<sup>43</sup> described a modified approach involving fulguration of the internal layers of the ureterocele to promote adhesion of these layers after collapse. This approach completely prevented the development of *de novo* VUR while achieving a decompression rate of 82%. The choice of endoscopic technique, however, seems to be important only for the balance between efficient decompression and avoidance of *de novo* reflux in the punctured moiety;<sup>43,45,46</sup> in general, the larger the incision, the more effective the decompression, but the higher the risk of *de novo* VUR. As discussed before, instead, the choice of decompression technique has little bearing on the treatment of associated VUR and for upper pole function improvement. The issue, therefore, is whether postoperative VUR and nonfunctioning upper poles mandate secondary open surgery, as has generally been considered so far.<sup>46</sup> Indeed, endoscopic decompression can be combined with endoscopic treatment of symptomatic VUR both in the punctured and associated moieties,<sup>10,43</sup> with a reported success rate after two injections ranging from 60% to 90%.<sup>38,47</sup> In one series, only 1 case out of 49 patients presenting with refluxing units treated by initial endoscopic puncture and subsequent endoscopic treatment of VUR required ureteric implantation.<sup>48</sup> Kajbafzadeh *et al.*<sup>43</sup> even proposed a preemptive endoscopic treatment of all ureters that showed signs of reflux on the micturating cystourethrography performed before endoscopic decompression, and reported a secondary surgery rate of 10% after 4.2 years' follow-up.

With regard to nonfunctioning or poorly functioning upper poles, Chertin *et al.*<sup>4</sup> reported no cases of hypertension and only 2 heminephrectomies for symptomatic infections out of 48 cases of nonfunctioning or poorly functioning upper poles left *in situ* after endoscopic decompression followed-up for a median period of 9 years.



**Figure 2** | Nephroureterectomy of a nonfunctioning upper pole through retroperitoneal laparoscopic approach. **a** | Renal scan showing a poorly functioning upper left pole on the left. **b** | Port sites and removed specimen.

### Upper tract surgery

The upper tract approach includes pyelopyelostomy in cases where the upper tract is functional or heminephrectomy in the case of nonfunctioning or poorly functioning upper moieties (which comprise ~70% of cases).<sup>3,4,18,29</sup> Hussmann *et al.*<sup>20,42</sup> showed that in patients with ectopic DSUs and no preoperative VUR, upper urinary tract surgery was significantly more effective than endoscopic decompression, and had an 80% chance of being the definitive treatment. It should be noted, however, that long-term follow-up studies reported a prevalence of *de novo* ipsilateral lower pole or contralateral reflux of 40–50% following upper tract surgery.<sup>6,20</sup>

Major concerns surrounding upper tract surgery are the need for a lumbotomy and the risk of iatrogenic injury to the lower pole vessels. The former can be avoided by using a laparoscopic approach for heminephrectomy (Figure 2), which, depending on experience, allows for results comparable to the standard open approach,<sup>49–51</sup> although it may still be a formidable procedure to undertake in infants.<sup>52,53</sup>

### Total reconstruction

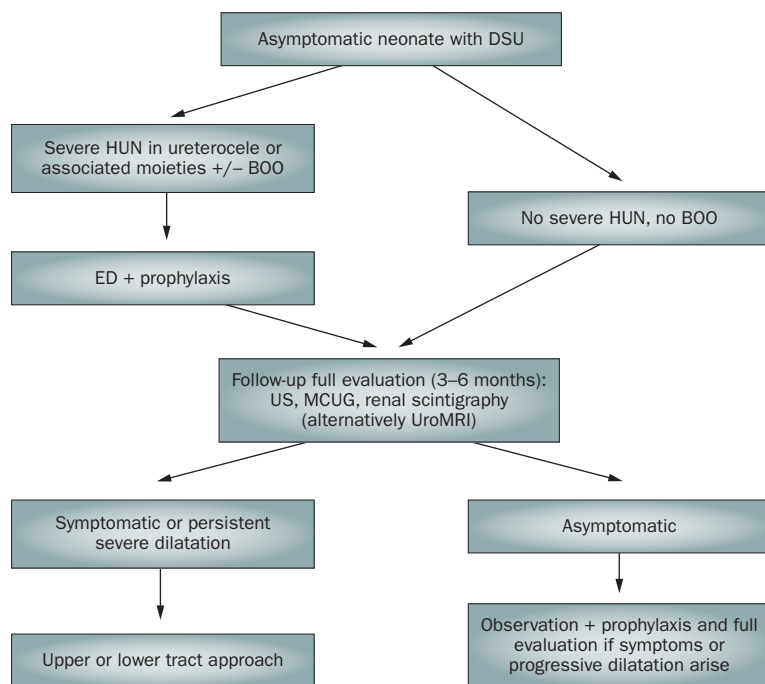
Previously, total reconstruction was seldom recommended in infants because of the risk of damaging the bladder, but with current standards of care it can now be performed safely at any age without clinical or urodynamic consequences for bladder function.<sup>8,9</sup> Although the role of total reconstruction for the prevention of urinary incontinence, as discussed above, is questionable, the procedure still represents the most effective and consistent approach for achieving single-stage cure of VUR.<sup>2,9,10</sup> It is of note, however, that even this approach is not 100% effective.<sup>2</sup> Beganovic *et al.*<sup>37</sup> reported persistent VUR in 13% of cases and a secondary surgery rate of 36% during long-term follow-up of 53 patients. Consequently, it has been recommended that this procedure be limited to cases of preoperative multiple or high-grade VUR.<sup>2,9,11</sup>

Attempts have been made to simplify total reconstruction. Gran *et al.*<sup>31</sup> proposed avoiding the upper tract approach and also reimplanting the ureter of

**Table 4** | Criteria for conservative ureterocele management and outcomes

Study	Patients (n)	Total study population (n)	Criteria	Follow-up (years)	Outcome
Shankar <i>et al.</i> (2001) <sup>12</sup>	14	52	Asymptomatic; lower pole less than grade III VUR; no lower pole non-refluxing HUN; no BOO; upper pole function <10%	8	No UTI or symptoms Dilatation: stable in 8 patients, improved in 6 VUR: resolved in 3 out of 4 patients
Merguerian <i>et al.</i> (2003) <sup>16</sup>	10	60	Minor upper pole dilatation; no evidence of obstruction on renal scan	5	No UTI or symptoms Dilatation: stable in 6 patients, resolved in 4
Coplen and Austin (2004) <sup>13</sup>	4	34	Multicystic dysplasia in the ureterocele moiety; no ureteric dilatation; less than grade III associated VUR; no BOO	1–5.5	1 UTI Ureterocele: collapsed in 2 patients VUR: resolved in 2 out of 3 patients All multicystic dysplastic moieties showed involution by 18 months
Han <i>et al.</i> (2005) <sup>14</sup>	11	ND	Good or no function at all in the ureterocele moiety; no evidence of high-grade obstruction on MAG3 scan (half-time <30 min)	3–4	Surgery required in 4 patients (3 UTIs, 1 progressive dilatation) Dilatation: stable in 1 patient, improved in 3, resolved in 3 VUR (grade III and IV): resolved in 5 out of 5 patients
Di Renna <i>et al.</i> (2006) <sup>15</sup>	6	ND	Asymptomatic; lower pole less than grade III VUR; no lower pole non-refluxing HUN; no BOO; upper pole function <10% or >10% but no obstruction	5	No UTI or symptoms Dilatation: stable in 1 patient, improved in 1, resolved in 4 VUR: stable in 1 patient, improved in 1, resolved in 4

Abbreviations: BOO, bladder outlet obstruction; HUN, hydroureteronephrosis; MAG3, <sup>99m</sup>Tc-mercaptoacetyltriglycine; ND, not determined; UTI, urinary tract infection; VUR, vesicoureteric reflux.



**Figure 3** | Algorithm for the management of duplex system ureteroceles in neonates. The presence of severe upper urinary tract dilatation, regardless of whether obstructive or refluxing, seems to be the most important risk factor for infections in asymptomatic neonates. Accordingly, endoscopic ureterocele decompression seems appropriate in the face of severe HUN. Asymptomatic neonates without massive HUN might instead commence antibiotic prophylaxis and undergo a comprehensive assessment at 3–6 months to assess anatomy and function of the different renoureteral units. Based on the results of this assessment and the clinical behavior during the period of observation, further conservative or surgical management is chosen according to particular criteria. Abbreviations: BOO, bladder outlet obstruction; DSU, duplex system ureterocele; ED, endoscopic decompression; HUN, hydroureteronephrosis; MCUG, micturating cystourethrography; UroMRI, MRI urography; US, ultrasonography.

the nonfunctioning pole; none of their 16 patients developed any problems during an average follow-up of 5 years. This result was confirmed by Wang *et al.*<sup>32</sup> Alternatively, ureteroureterostomy has been reported as a viable option, even in cases in which there is a marked discrepancy in diameter between the donor and recipient ureter.<sup>54,55</sup> Finally, others have suggested avoiding ureterocele excision and bladder-neck reconstruction and, instead, performing the whole procedure via an extravesical approach.<sup>16,36</sup>

**Conservative management**

This approach was initially considered by Rickwood in order to ascertain the natural history of antenatally diagnosed ureteroceles.<sup>12</sup> Conservative management can, however, also be a viable option for patients diagnosed with ureteroceles after presenting with a UTI.<sup>14</sup> The criteria proposed for nonoperative management vary according to the presenting clinical scenarios that are considered at lowest risk of complications, such as UTIs (Table 4). In general, the patients should not present with any bladder outlet obstruction, severe HUN of the ureterocele moiety or high-grade (over grade III) VUR.<sup>12,13,15</sup> Nevertheless, cases of successful conservative management have occasionally been reported in patients with severe ureterocele moiety HUN<sup>12</sup> or high-grade VUR.<sup>14</sup>

It has been suggested<sup>14,15</sup> that the scintigraphic drainage pattern might be more relevant for defining the presence of obstruction than the actual urinary tract dilatation, such that conservative management might also be extended to cases of ureteroceles that have demonstrable moiety function, provided that there is adequate drainage (half-time <30 min)<sup>14</sup> for the affected unit. Coplen and Austin,<sup>13</sup> however, have limited conservative



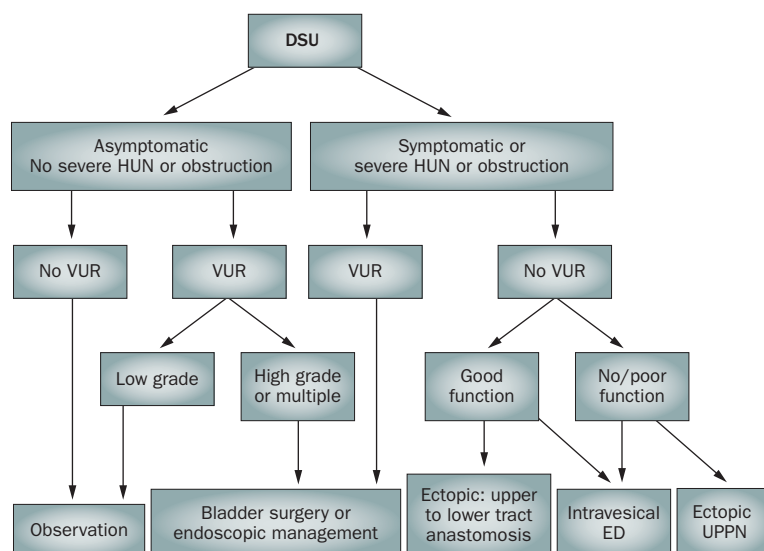
**Figure 4** | MRI urography of a prenatally detected hydronephrosis at 3 months. This approach facilitates the assessment of both anatomical and functional aspects of ureterocele, and might replace standard investigations. Image courtesy of the Pediatric Radiology Department of Robert Debré Children's Hospital, Paris, France.

management to patients with multicystic dysplasia in the upper pole, consistent with the principle that some form of surgery is otherwise necessary to ensure a completely unimpaired drainage.

### Conclusions

Owing to the lack of controlled trials, the limited number of case-control studies comparing different approaches,<sup>17,36,42</sup> and the limited knowledge of the natural history of the condition, an evidence-based choice of the initial management of DSU in neonates and infants is currently not possible. This Review shows that some of the classical arguments on which the choice of initial approach is usually based upon are indeed questionable, including the need for systematic ureterocele decompression to prevent UTIs<sup>17</sup> or improve renal function,<sup>23-25</sup> and the need for systematic bladder base/neck reconstruction to prevent incontinence.<sup>16,36</sup> Moreover, evidence does not show any increased morbidity owing to nonfunctioning upper poles left *in situ*,<sup>4,31,52</sup> and questions the causative relationship between VUR and febrile UTIs.<sup>20,33-36</sup>

In our view, treatment should generally be individualized, based on the results of a comprehensive preoperative evaluation and the clinical status of the patient. In the asymptomatic neonate that shows no evidence of bladder outlet obstruction or renal function abnormalities, the primary aim of treatment is to prevent UTIs. The presence of severe upper tract dilatation, regardless of whether obstructive or refluxing, seems to be the most important risk factor for infections. Accordingly, endoscopic ureterocele decompression seems appropriate in the face of severe HUN. Neonates without massive



**Figure 5** | Algorithm for the management of duplex system ureterocele after the first 3–6 months of life. Obstruction is considered to be the presence of non-refluxing dilatation of non-ureterocele-bearing moieties (especially of the lower pole)<sup>12,13</sup> or of an obstructive drainage pattern on diuretic renography.<sup>14,15</sup> Endoscopic management includes endoscopic decompression of ureterocele and endoscopic or conservative management of VUR. Abbreviations: DSU, duplex system ureterocele; ED, endoscopic decompression; HUN, hydroureteronephrosis; MCUG, micturating cystourethrography; UPPN, upper pole partial nephrectomy; VUR, vesicoureteral reflux.

HUN might instead commence antibiotic prophylaxis and undergo a comprehensive assessment at 3–6 months (Figures 3 and 4). Beyond this age, management can reasonably be chosen on the basis of the balance between the risk of infection or other untoward outcomes and the effectiveness of each single management option (Figure 5). In asymptomatic patients without bladder outlet obstruction, massive HUN or high-grade (over grade III) VUR, conservative management can be continued, as the risk of UTIs is low and the chance of spontaneous improvement is high.<sup>12,14,15</sup> In symptomatic patients, or in cases of high-grade VUR or massive HUN, surgical management is recommended.<sup>13,15,21</sup> In patients with high-grade HUN but no VUR, many studies have recommended differentiating the management on the basis of ureterocele position: endoscopic decompression is carried out for intravesical ureterocele whereas an upper tract approach is used for ectopic ureterocele.<sup>20,42</sup> Although we agree with the use of endoscopic decompression for small ureterocele (indeed, these are rarely associated with significant obstruction), ureterocele in neonates and infants are usually large, and differentiating them into either intravesical or ectopic is often difficult, and largely subjective.<sup>32</sup> In these cases, management choice might be based upon ureterocele moiety function, removing nonfunctioning upper poles and decompressing endoscopically those ureterocele with preserved function. DSUs with associated multiple or high-grade VUR represent the most challenging cases, and are also the most common. These

patients show minimal chance of spontaneous resolution of VUR after ureterocele decompression (either endoscopic or by heminephrectomy), irrespective of ureterocele position. Thus, if single-stage resolution of VUR is deemed necessary, some form of surgery to the bladder should be considered.<sup>2,19,49</sup> Alternatively, endoscopic decompression achieves definitive treatment in about half of these patients,<sup>2,9,16</sup> but if combined with conservative management of asymptomatic VUR<sup>4,32</sup> and minimally invasive management of symptomatic VUR,<sup>4,32,47</sup> the need for open surgery can be restricted to very selected cases.<sup>48</sup> Furthermore, no approach seems to be entirely free from some need for secondary surgery in the long term.<sup>6,20,37</sup>

Finally, as for primary VUR,<sup>56–58</sup> parental preference and costs should also be taken into account in the decision-making process, although data on these aspects are still limited.

#### Review criteria

References for this article were obtained by performing a thorough search via the databases MEDLINE/PubMed and EMBASE of English-language papers published over the past 15 years up to December 2008 using the MeSH terms “ureterocele”, “duplex system”, “hydronephrosis” and “pediatrics”. Results were pooled and a double review made, initially of titles and abstracts, and then of the full text of pertinent papers.

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