The Management of Ureteroceles

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Definitions and incidence

A ureterocele is a congenital cystic dilatation of the intravesical submucosal ureteral segment [26]. The terminology used to describe ureteroceles can be ambiguous and confusing so throughout this text the definitions suggested by Glassberg and colleagues in 1984 will be adhered to [12]. There are two types of ureterocele:

Intravesical - this is located entirely within the bladder and may be associated with either a single or a duplex system.

Ectopic - some of the ureterocele is situated permanently at the bladder neck or in the urethra. The orifice may be situated in the bladder, at the bladder neck or in the urethra.

Stephens in 1971 [24] further classified ureteroceles as stenotic, sphincteric, sphinctero-stenotic and cecoureteroceles. The cecoureterocele is mainly intravesical but has a blind extension of varying depth alongside the urethra. While these classifications improve our understanding of the condition they do not often influence therapy, with the possible exception of the cecoureterocele [23].

The exact incidence of ureteroceles is unknown but has been estimated to occur between 1:5000 and 1:12000 of pediatric admissions [19]. Brock and Kaplan stated that ureteroceles were not rare in urological practice being found in 1-2 % of patients undergoing cystoscopy [3]. Right and left sides are affected equally and there is a female to male ratio of at least 4:3 [27]. Complete duplication is present in 80-90 % of children with ureteroceles and approximately 15 % are bilateral [15].

Embryology

Although the aetiology of ureteroceles is unclear, there are a number of theories to their embryogenesis: Chwalla's membrane is an epithelial sheath that separates the Wolffian duct from the urogenital sinus. It is possible that a delayed rupture of this membrane would result in an aneurysmal dilataton of the distal ureter and stenosis of its orifice [21]. Mackie and Stephens considered ureteroceles to be a part of a generalised abnormality of the ureteric bud from the close association between ureterocele, ureteral duplication and renal dysplasia [18].

Tokunaka and colleagues suggested that a ureterocele is a segmental embryonal arrest of the most distal portion of the ureter with poorly developed muscle bundles, smaller muscle cells and an absence of thick myofilaments [26].

Pathology

When complete duplication is present the ureterocele is almost always associated with the upper pole, but Lima and Cavalcanti reported a ureterocele draining the lower pole [17].

Ectopic ureteroceles usually interfere considerably with the normal mechanics of the urinary tract and may cause any combination of reflux and/or obstruction involving all 4 renal units as well as bladder outlet obstruction [5]. In children most cases are associated with duplication and as the cranial ureteric bud misses the centre of the metanephric cap renal dysplasia will ensue. Caldamone and colleagues found that severe dysplasia or chronic pyelonephritis and fibrosis affected all of 35 specimens examined histologically [5]. Williams in 1958 [28] and Geringer et al in 1983 [9] found upper pole function in only 10-15 % of intravenous urograms. Although the ureterocele produces obstruction to the upper pole in the vast majority of cases, Bauer and Retik reported 5 children with unobstructed upper poles [2]. Messing and Henry reported stones in bilateral unobstructed intravesical ureteroceles associated with single systems [20]. In the majority of cases the dilatation of the lower pole is

secondary to reflux or obstruction by the ureterocele, but Androulakakis et al described 2 cases where the dilatation was a result of intrinsic pathology at the ureterovesical junction [1].

Presentation

In most large series at least 50 % of patients presented during the first year [5,7]. The commonest presentations were with urinary infection or septicemia, prolapse of ureterocele, abdominal mass, hematuria or abnormal voiding. In 1984 only 1 out of 58 patients reported by Caldamone et al [5] had an antenatal ultrasound diagnosis. This figure had risen to 9 % in 1989 [7] and in 1992 39 % of 41 patients reported by Rickwood et al [22] were diagnosed antenatally. It is likely that this increasing trend will continue and the majority of patients will present asymptomatically in this way. Management strategies may have to be reconsidered under these circumstances.

Investigation

The value of ultrasonography (US) is now well established in the investigation of the urinary tract and is ideally suited for demonstrating ureteroceles, the dilated ureter and in cases with duplication the hydronephrotic upper pole (Fig. 1). The appearance of a ureterocele has been described as a "cyst within a cyst" [13]. In cases where infection has occurred it may also demonstrate pus within the system (Fig. 2). On this basis ultrasound is the investigation of first choice and when combined with isotope renography the function and drainage of the various segments of the upper tracts can be accurately assessed (Fig. 3). This information is essential in choosing the correct treatment options for individual patients. Micturating cystourethrograms (MCUG) are necessary to demonstrate vesico-ureteric reflux and are also helpful in excluding bladder outflow obstruction in patients with ectopic ureteroceles (Fig. 4).

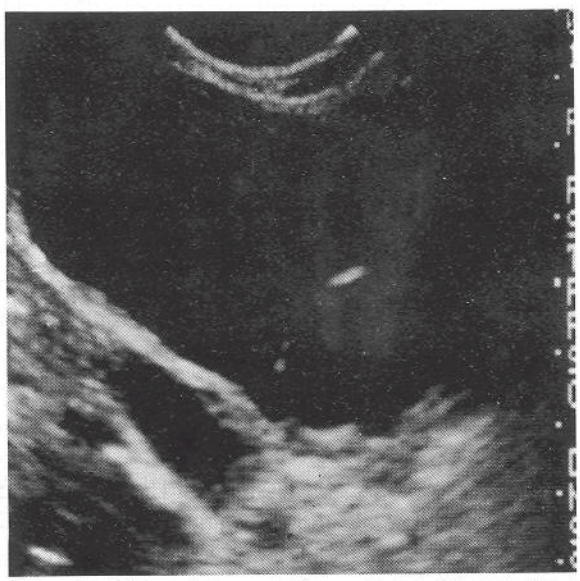


Figure 1. Ultrasound demonstrating a dilated ureter entering a ureterocele within the bladder.

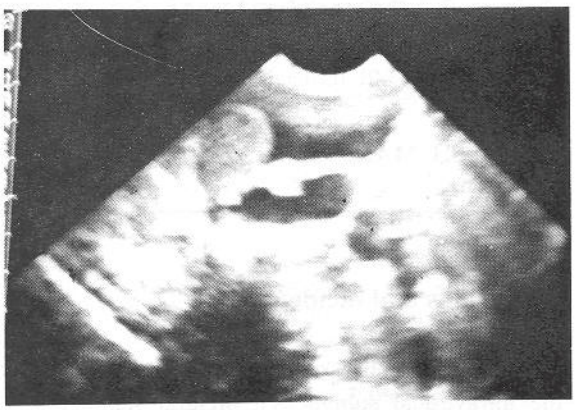


Figure 2. Ultrasound demonstrating echogenic material (pus) within the ureterocele.

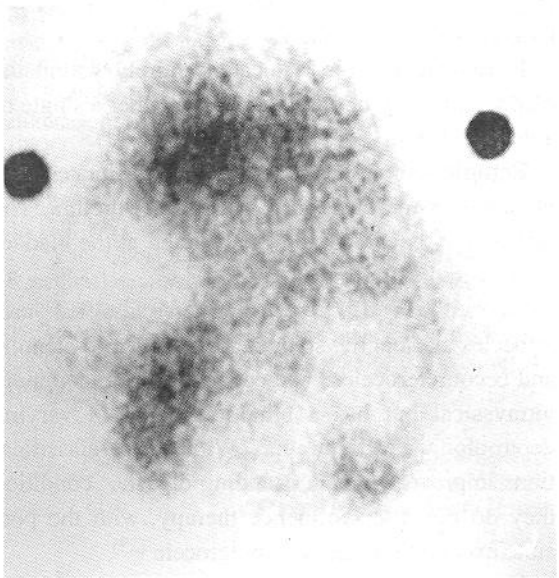
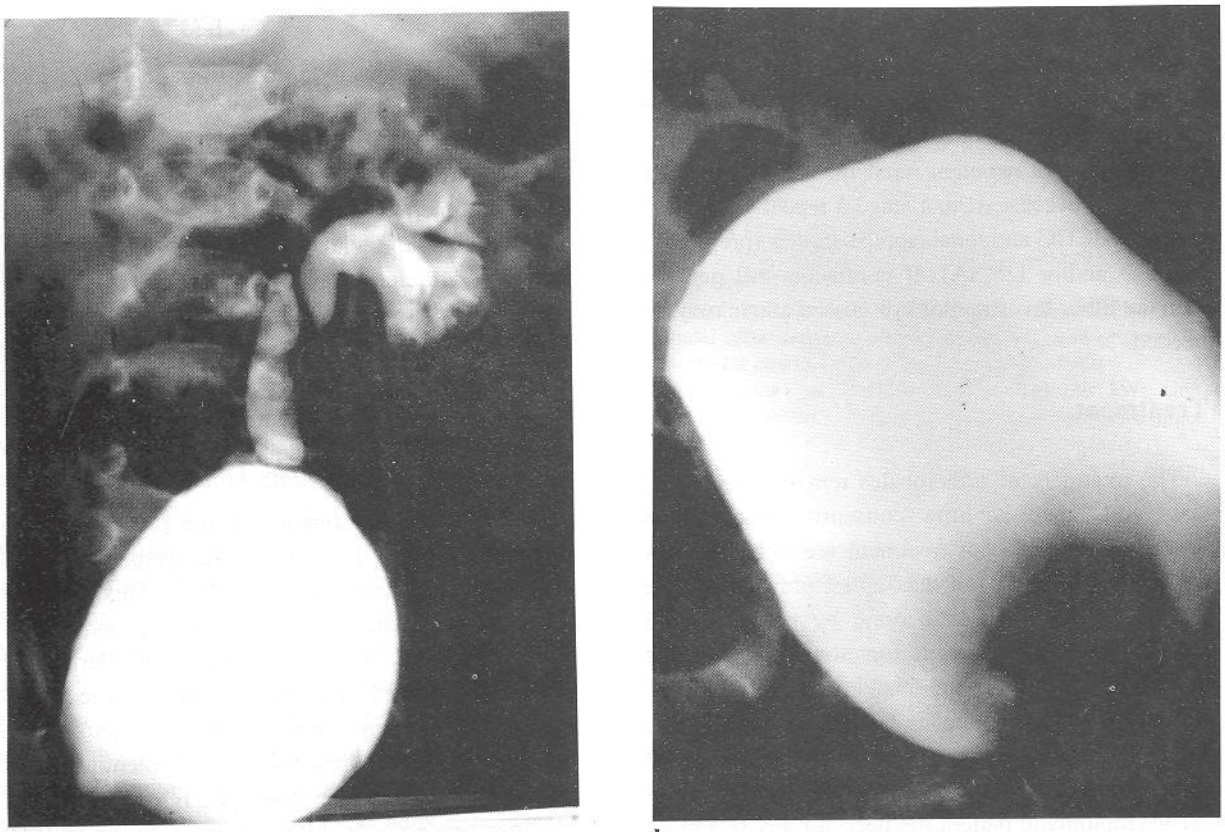


Figure 3. Renogram demonstrating an upper pole defect caused by a non-functioning upper moiety.



a b

Figure 4. MCUG demonstrating reflux into the ipsilateral lower pole (**a**) and the ureterocele on the lateral view (**b**).

It is possible to miss a ureterocele on MCUG by overfilling the bladder and causing it to collapse or simply by obscuring it by the presence of dense contrast in the bladder. Further studies are generally not required unless there is still doubt about the diagnosis or in deciding from which side the ureterocele is arising, a situation that occurs in a small number of cases. Intravenous urography (IVU) can then be helpful. Typical features on IVU include: the "drooping lily" sign of the lower pole collecting system as it is pushed downwards and laterally by the hydronephrotic upper pole; the "rim sign" as a result of opacification of the thinned parenchyma of the upper pole; the large intravesical filling defect of the ureterocele (Fig. 5). If confusion still remains cystoscopy may be required but the findings can be difficult to interpret in a small bladder with a large ureterocele.

The advent of increasing numbers of patients with asymptomatic presentations following antenatal diagnosis necessitates protocols for their investigation. Many are available and the following is just one example: Postnatal US at 3-5 days of age, if the diagnosis is confirmed and there is no evidence of

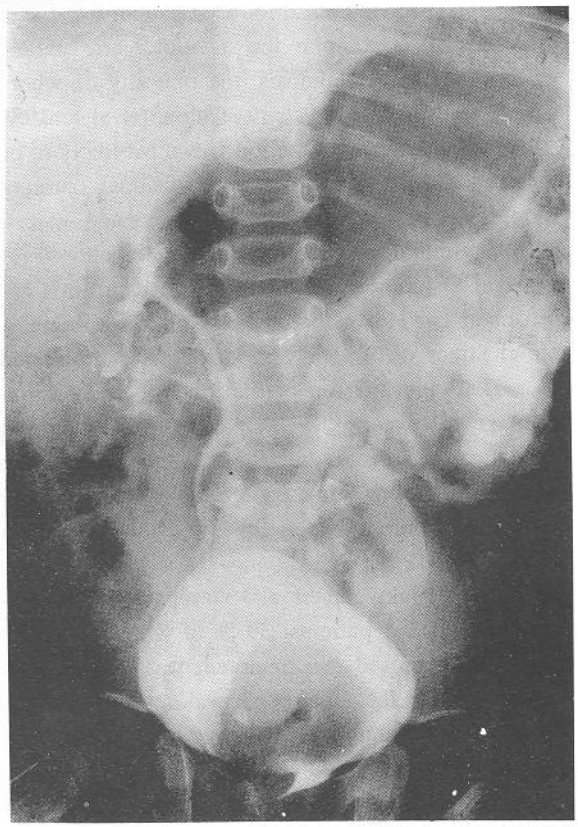


Figure 5. IVU demonstrating the typical features described in the text.

bladder outflow obstruction prophylactic antibiotics are started (Trimethoprim 2 mg/kg once a day) and further investigations are deferred until the initial period of transient neonatal renal function has passed at around 6 weeks. At that stage a repeat US is performed, MCUG and renal isotope studies (preferably MAG 3 and/or DMSA). If confusion still persists then the other investigations discussed above may be needed.

Treatment

The treatment of ureteroceles remains controversial and there is little consensus amongst the "experts". The aims of treatment are to relieve obstruction and stasis and thus either prevent or treat infection. This should be done by preserving all renal tissue with reasonable functional and avoiding complications within the bladder that could contribute to later incontinence or outflow obstruction. When the patient has presented symptomatically there is little debate about the need for surgery but in the asymptomatic patient the need for any treatment is questioned by some [22]. The treatment obviously needs to be individualised for each patient [7].

Let us first examine the asymptomatic patient with an antenatal diagnosis, the majority of whom will have duplicated systems. Caione et al in 1989 [4] reported that renal function was most likely to be preserved in this group and excisional surgery should be avoided. This has led to renewed interest in transurethral incision of the ureterocele [10,25]. Etker et al reported their experience using this approach at The Hospitals for Sick Children, Great Ormond Street [8]. A total of 23 patients were treated by endoscopic incision of the ureterocele at a median age of 4 weeks. Twelve patients (52 %) did not need further surgery, but the rest did need upper pole hemi-nephrectomy for urinary infection with residual dilatation though this was delayed for an average period of 1 year.

In 1992 Rickwood et al [22] reported their experience with 41 patients, 39 % of whom were antenatally diagnosed. No treatment was offered to 8 patients where the ureterocele was intravesical and there was no dilatation of the upper tracts except that pole obstructed by the ureterocele. At a mean follow up of 2.3 years no problems have been encountered but with this short follow up one must be cautious as in some large series over 26 % of patients presented with sepsis when they were older than 3 years [7]. However it does demonstrate that urgent neonatal surgery is not required.

When function is preserved other options include pyelopyelostomy with aspiration of the remaining ureter and ureterocele or an excision of the ureterocele with reimplantation of both ureters. In the author's experience lower tract surgery is very difficult in the small infant and in general should be avoided and the best temporizing option is endoscopic incision with delayed secondary surgery when necessary.

When upper pole function is not preserved there seems to be little point in saving dysplastic tissue and excisional surgery is indicated [5]. This principle would apply equally to asymptomatic and symptomatic patients. In 1979 Kroovand and Perlmutter [16] recommended simultaneous upper pole heminephroureterectomy, extravesical excision of the ureterocele (to avoid the tedious and potentially dangerous intravesical dissection) and reimplantation of the lower pole ureter. Johnston and Johnson in 1969 advocated excision of the ureterocele, reimplantation of the lower pole ureter and subsequent upper pole nephrectomy [14]. Following this approach Brock and Kaplan reported a successful outcome in 11 out of 16 patients; three had persistent ipsilateral vesicoureteric reflux, one patient developed a stone on a bladder suture and one had a large bladder diverticulum [3]. There is little doubt that this aggressive approach is not without difficulty or hazard, complications including persistent reflux, obstruction, infection, incontinence or other micturition problems [29].

Because of these difficulties it has been advocated that the initial treatment should be upper pole heminephrectomy with aspiration of the ureterocele via the distal ureteric remnant. It is also claimed that this approach makes for easier excision of the ureterocele should it become necessary. In 1983 King et al [15] found that only 10 out of 39 patients (26 %) treated by this approach needed subsequent surgery. More recently Rickwood et al [22] reported that only 9 % of their patients treated in this way needed further surgery.

However Caldamone et al in 1984 [5] and Decter et al in 1989 [7] found that 50 % and 54 % of their patients respectively needed further surgery for per-

sistent reflux or problems with the ureterocele. In this author's opinion these high reoperation rates are not an argument against the more conservative surgical approach. At least 50 % of patients will need no further surgery and where surgery is required it is not complicated by the previous hemi-nephrectomy and may in fact be made easier.

Specific therapeutic measures may have to be considered in certain cases. When there is a prolapse of the ureterocele initial manual reduction should be attempted with needle aspiration if it is not successful [11]. If this fails a simple external incision of the ureterocele should allow subsequent reduction [6]. When a patient presents with septicemia secondary to pyoureteronephrosis intravenous broad-spectrum antibiotics are required and the patient should be fully resuscitated before surgical intervention is considered. A number of options are available including ultrasound guided percutaneous nephrostomy drainage, endoscopic incision of the ureterocele to establish decompression, but in the author's experience if a DMSA scan reveals a non-functioning pole an emergency hemi-nephroureterectomy is a safe and potentially a definitive procedure.

Specific mention must be made of the ureterocele associated with the single system in childhood. Renal function is commonly preserved and a simple endoscopic incision of the ureterocele should relieve obstruction and reflux does not commonly occur [30]. When function is poor a nephroureterectomy without ureterocele excision is usually all that is required.

Finally it is important to discuss some specific postoperative complications of ureterocele surgery: Bladder outlet obstruction may result from a persisting tense ureterocele acting as a ball-valve on the bladder neck or from a ureterocele with a poor muscle backing which acts as a diverticulum compressing the bladder neck area.

Patients who have undergone a previous ureterocele excision may suffer bladder outlet obstruction as a result of a flap-valve produced by a residual lip of ureterocele tissue at the bladder neck. Transurethral excision of this lip is usually all that is required in these patients. Although many controversies still exist there is little doubt that whatever treatment modality is chosen the overall prognosis is very good.

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