A 10-year old boy presented with a six-month history of difficulty passing urine as well as dribbling and incontinence. He had been treated by his family doctor for urinary tract infections however his symptoms had recently deteriorated. He was otherwise well with no prior medical history of note. An ultrasound of the kidneys demonstrated some scarring of the upper pole of the left kidney but was otherwise normal with no evidence of a duplicated system. Pelvic ultrasonography demonstrated bilateral large ureteroceles, which partially covered the bladder outlet (Fig. 1). A micturating cystourethrogram (MCUG) was then performed. This again demonstrated bilateral ureteroceles. It was noted that towards the end of micturition, both ureteroceles prolapsed into the urethra and caused some obstruction to flow leading to incomplete bladder emptying (Fig. 2).

An isotope DMSA scan (not shown) demonstrated evidence of left-sided reflux nephropathy with percentage uptake of 32% on this side. Cystoscopy confirmed the presence of large ureteroceles bilaterally, with prolapse into the bladder neck and proximal urethra. Single ureteric orifices were noted bilaterally, which were located intravesically. Endoscopic puncture of bilateral ureteroceles was performed, with frank pus draining from both puncture sites. The patient was commenced on prophylactic I-V antibiotics and discharged home well 4 days post-operatively. At one-month follow-up, there was complete resolution of symptoms with radiological evidence of collapse of both ureteroceles on ultrasound studies.

**Discussion**

A ureterocele refers to a cystic dilatation of the distal ureter protruding into the bladder lumen. This malformation is most commonly diagnosed in the paediatric population...
tion for two reasons: firstly, due to prenatal ultrasound screening for congenital anomalies and secondly, due to the high incidence of postnatal urinary tract infections, resulting in ultrasonography of the renal tract as a first-line investigation. The incidence is reported to be between 1 in 5000 to 1 in 12000 of paediatric hospital admissions (1), however a number of autopsy studies have suggested a much higher incidence of up to 1 in 500 (2). It occurs 4-6 times more frequently in girls and is observed almost exclusively in the Caucasian population (3). Ureteroceles have no predilection for side and 10% of cases show bilateralism (4). A number of classification systems exist based on varied anatomical and pathological criteria. However, the most widely accepted and most frequently used system has been established by the American Academy of Paediatrics (5), which classifies ureteroceles contained entirely within the bladder (i.e. ureteric orifice and ureterocele itself) as intravesical or orthotopic, whereas those with some portion of the ureterocele permanently located at the bladder neck (or rarely urethra) as ectopic. Up to 80% of paediatric ureteroceles occur with ureteric duplication anomalies and are termed duplex-system ureteroceles, with 60-80% of these having an ectopic drainage orifice (6). In contrast to the majority of paediatric ureteroceles, those presenting in adulthood are typically single-system ureteroceles, which insert in a normal position in the trigone of the bladder, with only mild dilatation of the distal ureter. Although single-system ureteroceles usually manifest in adulthood, they have been reported with increasing frequency in recent years amongst urologists and radiologists in paediatric practice (7). Single-system ureteroceles are distinguishable clinically from the more common duplex-system ureterocele by their frequent occurrence in male children (in direct contrast to female predominance in duplex-system ureteroceles) and their association with multicystic dysplastic kidney (7).

The aetiology of ureterocele has never been definitively established. A number of theories exist and most conclude that the underlying abnormality is mucosal in origin and ureteroceles, in particular ectopic ones, involve a defect of the muscle coat of the affected ureter and often a defect in the bladder wall. Moreover, it is thought that the ureteroceles associated with single systems are not all congenital and may be acquired in adulthood (6). However no one theory satisfactorily explains the different types of ureterocele and it is thought unlikely that the underlying aetiology has any impact on clinical practice and management of ureterocele.

The clinical presentation is varied. The advent of routine prenatal ultrasonography as part of obstetric care in the developed world has significantly affected the mode of presentation as well as the age at presentation of paediatric ureteroceles, with the number of neonates with prenatally detected ureteroceles having increased from 2% to 28% over the last two decades (8). Furthermore, prenatal diagnosis of ureterocele represents 15% of all prenatally diagnosed duplex kidneys (9). Prior to this, almost all children presented in early childhood with symptoms and signs of urinary tract infection, and overall, this is still the most common mode of presentation in children of both sexes (10). However, a number of other presentations are reported (11). Bladder outlet obstruction is extremely rare and may occur secondary to a prolapsing ureterocele at the bladder neck or proximal urethra. In this case, we have described outlet obstruction occurring secondary to both ureteroceles extending into the proximal urethra, likely resulting in a ball-valve effect. A prolapsed ectopic ureterocele in a female child may rarely present as an inter-labial mass. Rarely, children with ureteroceles may present with haematuria after minimal trauma (9). Other symptoms may be more non-specific and include failure to thrive or vague abdominal pain (4). Urinary incontinence may occur due to interference with normal sphincter function at or distal to the bladder neck by an ectopic ureterocele (11). In our patient’s case, it was felt that the large ureteroceles prolapsing into the bladder neck as demonstrated on ultrasound, MCU and at cystoscopy, interfered with sphincter function in this way to cause a significant degree of urinary incontinence. The cystic distal ureter likely allowed stasis of urine and promoted the development of infection, resulting in the pus formation which was drained at endoscopic puncture. Endoscopic puncture has been found to be very successful at achieving decompression (91%) of a ureterocele (12). Some patients may require a second procedure, so follow up is required.

References