Clinics in Diagnostic Imaging (61)

K B J Sheah, S K H Yip, V T Joseph

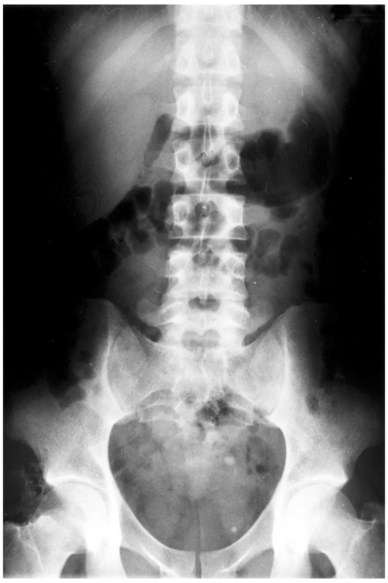

Fig. 1a Control abdominal radiograph.

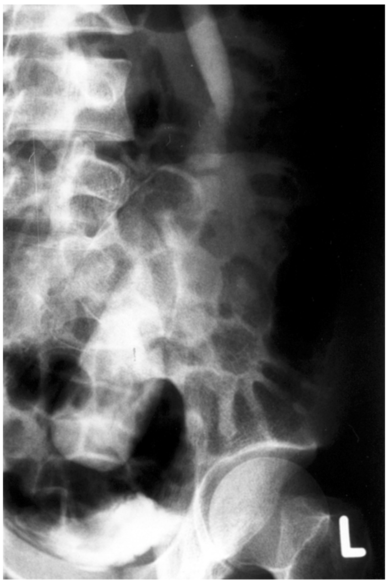

Fig. 1b Coned oblique IVU radiograph of left lower ureter.

Fig. 1c IVU post-micturition radiograph.

CASE PRESENTATION

A 27-year-old man presented with sudden onset of sharp left loin accompanied by nausea and vomiting. The pain was worse on movement and was non-radiating. He was afebrile and bowel motions were normal. On retrospective review, he had been experiencing this pain intermittently for nearly four years, during which he required hospitalisation for pain control on one occasion. Physical examination during the current admission was unremarkable.

He was discharged after adequate pain control and rehydration. Intravenous urography (IVU) was subsequently performed (Figs. 1a-c). What does the IVU show? What is the diagnosis?

Department of Diagnostic Radiology
Singapore General Hospital
Outram Road
Singapore 169608

K B J Sheah, MBBS
Medical Officer

Department of Urology

S K H Yip, FRCS, FAMS, FHKAM
Consultant

V T Joseph, FRACS, FAMS
Visiting Senior Consultant

Correspondence to:
Dr Sidney K H Yip
Tel: (65) 321 4693
Fax: (65) 227 3787
Email: gurykh@sgh.gov.sg

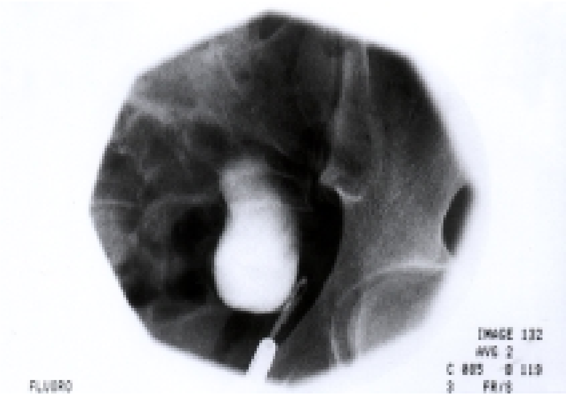

Fig. 2 On-table retrograde pyelogram shows a S-shaped stenotic ureter and a grossly dilated ureter above stenotic segment.

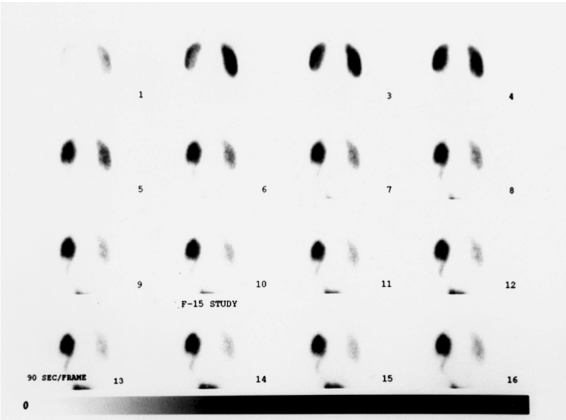

Fig. 3a DTPA scintiscans show persistent hold-up of tracer at the left lower ureter, suggestive of obstruction.

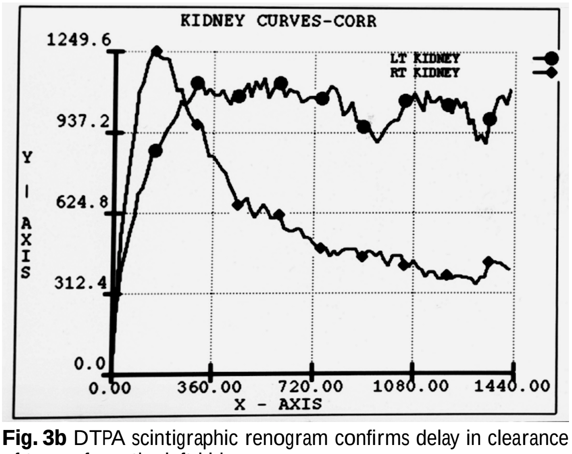

Fig. 3b DTPA scintigraphic renogram confirms delay in clearance of tracer from the left kidney.

IMAGE INTERPRETATION

The initial control radiograph shows a small rounded calcified opacity projected over the left hemipelvis (Fig. 1a). IVU confirmed that the opacity was located in a dilated lower left ureter (Figs. 1b-c). The other smaller left-sided opacity was located outside the urinary tract and represented a phlebolith. The ureter was grossly dilated and there was a stricture at its lower end. Moderate left hydronephrosis was present.

DIAGNOSIS

Left primary megaureter with lower ureteric stone.

Fig. 4 Post-operative IVU shows resolution of obstruction and absence of hold-up.

CLINICAL COURSE

The patient underwent cystoscopy, during which ureterocoele was excluded. On-table retrograde pyelogram showed a 3 cm stenotic segment in the left lower ureter, just distal to the grossly-dilated portion of the ureter (Fig. 2). Balloon dilatation and double pigtail ureteric stenting were performed. The internal stent was removed after four weeks. However, he rapidly developed repeated episodes of left loin pain. A ^{99m}Tc DTPA renal scan was then performed, revealing preservation of left renal function but evidence of isotope hold-up at the lower ureter, suggestive of persistent obstruction (Figs. 3a-b).

Subsequently, the patient underwent an open surgical exploration. The lower ureter was mobilised through an anterior cystostomy, starting from the ureteric orifice and continuing proximally. The stenosed 3 cm segment was resected. The ureteric stone was located at the opened proximal ureter and retrieved using a basket via a flexible ureteroscope. The ureter was pulled through a submucosal tunnel to create an anti-reflux mechanism without trimming or tubularisation. A new ureteric opening was constructed by joining the transected ureter to the mucosal surface of the bladder. A ureteric stent was then inserted.

Follow-up ultrasonography (US) showed complete resolution of dilatation. The patient remained well

Fig. 5 Micturating cystogram of another young man with neurogenic bladder shows gross secondary vesico-ureteric reflux leading to dilatation of the right ureter and pelvicalyceal system.

after removal of ureteric stent. Post-operative (four months) IVU demonstrated complete resolution of the obstruction, without any contrast hold-up in the ureter (Fig. 4).

DISCUSSION

Megaureter consists of a spectrum of anomalies involving the ureter. The international classification, mechanism and causes are outlined in the Table. Issues that warrant consideration are (1) how to differentiate obstructive megaureter from non-obstructive megaureter, and (2) when to intervene. It is the obstructive form of megaureter that may give rise to complications, which warrants surgical intervention. In this context, the current case of primary obstructed megaureter is presented to illustrate these principles.

Primary obstructed megaureter refers to a ureter that is obstructed and dilated disproportionally to the rest of the urinary tract[1]. The various causes of obstruction include congenital ureteral strictures, ureteral valves, ectopic ureter at the bladder neck, or a distal adynamic segment. Specifically, a distal adynamic segment is a disorder arising from failure of propagation of the peristaltic wave through a short extravesical adynamic segment of the distal ureter near the ureterovesical junction (3-4 cm), leading to functional obstruction that causes the proximal ureter to dilate.

Megaureter is more common in male than in female (ratio 2-5:1) patients, and the left side is more often affected than the right[1]. The abnormality is bilateral in 20-50% of cases. In general, the condition is diagnosed radiologically. In neonates and children, urinary tract infection or urinary symptoms often prompts further imaging studies. In contrast, the diagnosis of these congenital abnormalities in adulthood depends largely on the index of suspicion and threshold for radiological imaging. Adult megaureter is rare, as evidenced by the observation that out of 24,000 new adult genito-urinary cases over a period of 12 years, only 11 patients were diagnosed with the condition[2]. They often present with loin pain, recurrent urinary tract infection, and haematuria. Of these, pain is the most common presenting symptom[2].

The abnormality is often suspected when hydronephrosis and hydroureter are detected by renal US. The degree of collecting system dilatation may be graded subjectively and is more useful as a tool for monitoring, rather than diagnosing the severity of megaureter. However, US alone cannot elucidate the physiology and function of the dilated system[3]. IVU is the preferred modality for further investigation. In mild cases, there is a 2-3 cm fusiform dilatation of the lower ureter just proximal to the tapered extravesical distal segment[4]. With increasing severity, the ureter dilates more proximally to involve the pelvicalyceal system. In extreme cases, there is significant hydronephrosis, wasting of the renal parenchyma and impaired renal function. Fluoroscopy during IVU may demonstrate abnormal peristalsis in the proximally dilated ureter, where in the walls are unable to coapt.

Differential diagnoses include vesico-ureteric reflux, with or without underlying voiding dysfunction. To rule out reflux, micturating cystourethrography (MCU) may be performed. The quality of the bladder and urethra can also be assessed, as neurogenic bladder or outlet obstruction are common causes of secondary megaureter[5,6] (Fig. 5). Retrograde pyelography is a useful adjunctive technique employed in patients for whom surgical correction is contemplated. The initial cystoscopy prior to ureteral catheterisation is useful to rule out abnormalities in the trigone or ureteral openings[2]. Confirmation of the normal calibre of the aperistaltic segment further reinforces the diagnosis.

These investigations are important to identify primary obstructive megaureter due to a distal adynamic segment, or secondary obstructive megaureter due to valves or ureterocoele[7]. Hence, it is of utmost importance to elucidate that the presence of a dilated ureter may not necessarily imply significant functional obstruction. When the above-mentioned investigations have ruled out reflux, a non-obstructed non-refluxing

Table I Classification of megaureter

Mechanism		Causes
Reflux	Primary	Congenital refluxing megaureter Prune belly syndrome
	Secondary	Posterior urethral valves Neurogenic bladder
Obstructed megaureter	Primary	Adynamic segment Functional obstruction
	Secondary	Posterior urethral valves Neurogenic bladder Retroperitoneal scarring or tumour
Non-refluxing,	Primary	Idiopathic
non-obstructed megaureter	Secondary	Diabetes insipidus Infection Residual post-obstructive dilatation

megaureter needs to be differentiated from obstructive megaureter. In this regard, a diuretic renogram with ^{99m}technetium-diethylene-triaminepentaacetic acid (^{99m}Tc-DTPA) would be a better option[8]. The diuretic renogram of frusemide-assisted scintiscan measures the ability of a dilated system to empty following a diuretic challenge. It should be performed with the patient well-hydrated. Following intravenous administration of ^{99m}Tc-DTPA, initial images representing the angiographic phase provide an estimate of renal blood flow. After three to four minutes, renal perfusion films are obtained. Subsequently, images are taken at regular intervals to generate a renogram curve. Once the system is filled with ^{99m}Tc-DTPA, frusemide is injected and several films are taken. The half-time clearance of ^{99m}Tc-DTPA after administration of frusemide is under 15 minutes. Obstruction becomes more likely if clearance is prolonged more than 20 minutes (Fig. 3b).

The findings of the diuretic renogram can be affected by the degree of hydration of the patient, which in turn affects the clearance of the tracer[8]. In addition, presence of significant residual urine in the bladder also influences the result. Hence, the patient should be catheterised to ensure adequate bladder emptying. This is pertinent if the dilatation of the ureter is secondary to high bladder pressure. Lastly, poor renal function will jeopardise adequate excretion of ^{99m}Tc-DTPA to elucidate the cause of megaureter[7]. Despite these pitfalls, the diuretic renogram is a simple non-invasive test, which provides an accurate estimate of renal function. It can also be used to monitor patients serially.

Pressure perfusion study is another modality in the evaluation of obstruction in a dilated system[9]. The method involves a percutaneous renal puncture followed by an infusion of normal saline at 10 ml/min. The pressures within the renal system and the bladder are measured. A pressure differential of up to 15 cm H_20 is considered to be within normal limits. A higher value is suggestive of significant obstruction. However, this test is not often performed at our institution due to logistical difficulty, its invasive nature, and the lack of renal function information. Moreover, the pressure perfusion study is unsuitable for serial monitoring.

While uncomplicated megaureter does not require any specific treatment, some patients with recurrent urinary tract infections will require long-term prophylactic antibiotics[10]. Patients with recurrent symptoms (pain, stones, or infections) or significant renal impairment warrant a careful consideration of surgical correction. The commonest indication for surgical intervention is a history of recurrent admissions for loin pain, stones and impending renal impairment. In these cases, the operation of choice is excision of the affected ureteric segment, and re-implantation of the ureter into the bladder with an anti-reflux technique[2]. The objective of surgery is to provide a short route and direct good urine flow mainly by gravity, even in the absence of ureteric peristalsis. Ureteric tailoring is sometimes employed to provide a good fit for re-implantation and theoretically, this technique allows the ureter walls to coapt and promotes peristalsis[1]. To prevent reflux, the ureter is tunneled into the bladder wall[11]. The objective is to use intravesical pressure to close the ureteric tunnel when the bladder is full.

The surgical repair of megaureter is well described in children. The absence of urinary tract infection, progressive resolution of the size of ureteric dilatation by further imaging, and general improvement in condition are the basic criteria for successful outcome[11]. Owing to the relative rarity of this condition in adults, not many audited studies of the surgical outcomes are available. However, functional outcome are generally good with resolution of symptoms on subsequent follow up[12]. In this regard, the availability of ultrasonography in the clinic setting allows readily available monitoring of the patient on a regular basis.

REFERENCES

1. Atala A, Keating MA. Megaureter. In: PC Walsh, AB Retik, ED Vaughan, AJ Wein, eds. Campbell's Urology. 7th ed. Philadelphia: WB Saunders, 1998; 1897-906.
2. Hamilton S, Fitzpatrick JM: Primary non-obstructive megaureter in adults. Clin Radiol 1987; 38:181-5.
3. Platt JF. Urinary obstruction. Radiol Clin North Am 1996; 34:1113-29.
4. Pfister RC, McLoughlin AP III, Leadbetter WF. Radiological evaluation of primary megaureter. Radiology 1971; 99:503-10.
5. Davidson AJ, Hartman DS: The dilated pelvicalyceal system. In: Davidson AJ, Hartman DS, eds. Radiology of the Kidney and Urinary Tract. New York: WB Saunders, 1994; 596-600.

6. Blickman JG, Lebowitz RL. The coexistence of primary megaureter and reflux. AJR, 1984; 143:1053-7.
7. Norman N H. The wide ureter. In: Gillenwater JY, Grayhack JT, Howards SS, Duckett JW, eds. Adult and Pediatric Urology. St Louis: Mosby Year Book, 1991; 1863-85.
8. Sarkar SD. Diuretic renography: concepts and controversies. Urol Radiol 1992; 14:79-84.
9. Whitaker RH. An evaluation of 170 diagnostic pressure flow studies of the upper urinary tract. J Urol 1979; 121:602
10. Baskin LS, Zderic SA, Synder HM, Duckett JW. Primary dilated megaureter: long-term follow-up. J Urol 1994; 152:618-21.
11. Perovic S. Surgical treatment of megaureters using detrusor tunneling extravesical ureteroneocystostomy. J Urol 1994; 152:622-5.
12. Hanna ML, Wyatt JK. Primary obstructive megaureter in adults. J Urol 1975; 113:328-34.

ABSTRACT

Megaureter is infrequently diagnosed in adulthood. A 27-year-old man who presented with recurrent left loin pain was found to have megaureter on intravenous urography. His symptoms did not resolve after balloon dilatation of the 3 cm stenotic aperistaltic segment of the lower ureter. He eventually underwent ureteric re-implantation with satisfactory symptomatic relief. The resolution of obstruction was demonstrated radiologically. The role of imaging in the diagnosis and surveillance of megaureter, as well as indications for intervention, are discussed.

Keywords: Megaureter, urography, hydronephrosis, urinary tract, ureteric stone