Megaureter Management - Five-year Retrospective study

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Abstract:

**Purpose**: Management of patients with megaureter (MGU) and differentiating nonobstructive from obstructive variants, and better defining the indications for surgery, remains one of the more challenging dilemmas in Urology. We reviewed our experience in treating megaureter according to the diverse aetiologies of ureteral dilation.

**Materials and methods**: 15 patients of megaureter were chosen in the period of five years duration (Refluxing 7 cases, obstructed 6 cases, and non-refluxing non-obstructed 2 cases). Male to female ratio was 9:6. Age (1 month-35 years). Localization was: Lt side 57%, Rt. Side 36%, Bilateral 7%.

**Results**: Good results achieved in 87% of patients.

**Conclusions**: Precise definition of the cause of the dilation and proper patient selection may avoid complications in the reconstructive surgery.

**Keywords**: megaureter, obstructive, refluxing, treatment.

1. **INTRODUCTION**

The normal ureteral diameter in children is rarely greater than 5 mm and ureter wider than 7 mm can be considered megaureter (1). The term megaureter could be applied to any dilated or “big” (mega) ureter. The Paediatric Urology Society in 1976 adopted a standard nomenclature for categorizing the megal(o)ureter that had its basis in the diverse aetiologies of ureteral dilation. The dilated ureter or MGU can be classified into one of four groups based on the cause of the dilation (1) Refluxing (2) Obstructed (3) Both refluxing and obstructed and (4) Both non-refluxing and non-obstructed. Further subdivision into primary or secondary causes assume additional importance for obvious reasons. A thorough evaluation of the entire urinary tract is required in every case because therapeutic recommendations depend on proper categorization. Obstruction at the uretero-vesical junction is 4 times more common in boys than girls. It is often bilateral but usually asymmetric. The left ureter is slightly more often involved than the right (2). Close observation either at operation or by fluoroscopy reveals a failure of the distal ureter to transmit the normal peristaltic wave, resulting in a functional obstruction. Moreover, on fluoroscopy, retrograde peristalsis is seen which transmits abnormal pressures up to the kidney, resulting in calyceal dilation. Histologic findings include an excess of circular muscle, fibres and collagen in the distal ureter which may account for the problem (2).

2. **MATERIALS AND METHODS**

Fifteen patients with megaureter were chosen in the period of five years duration. Our cases of megaureters, whether being primary or secondary type, have been separated into three major categories: refluxing (7 cases), obstructed (6 cases), and non-refluxing non-obstructed (2 cases).

Male to female ratio was 9:6. Age ranged from one month to 35 years, median 11 years. Localization was: Lt Side 57% Rt. Side 36% Bilateral 7%. Patients were investigated during physical examination blood and urine analysis, ultrasonography, urography, voiding cystogram, Renal Isotopes scan and urodynamic study.

1) **Refluxing megaureter**: 7 patients

A- Primary

-(Primary refluxing megaureter) 2 patients

B- Secondary

-(Bladder outlet obstruction) 5 patients

-Obstructive Ureterocoele 1 patient

-Urethral stricture 1 patient

-post-urethral valves 2 patients

-Neuropathic bladder 1 patient

2) **Obstructed megaureters**: 6 patients

A- Primary:

-Intrinsic obstruction(Ureterocoele) 2 patients

-Stenosis 1 patient

-Adynamic segment 1 patient

B- Secondary:

- Neuropathic bladder 1 patient

-Retroperitoneal scarring 1 patient

3) **Non-refluxing non-obstructed**: 2 patients

A. Primary:

-Non-refluxing non-obstructed 1 patient

B. Secondary:

-Dilatation remaining after relief of distal obstruction. 1 patient
3. TREATMENT

Treatment ranged between remodelling of the ureter and reimplantation and was managed expectantly regarding the aetiology. Ureteral reimplantation (Politano-Leadbetter technique) with excision of the distal ureter was performed and because of the excessive dilatation of the ureter, ureteral tapering was necessary. In some cases, nephroureterectomy in dysplastic kidney and non-functioning upper moiety of the kidney with ureterectomy of refluxing tortuous severely dilated ureter. Ablation of posterior urethral valves was performed. Medical treatment with anticholinergic drugs was offered to those cases with neuropathic bladder.

4. RESULTS

Good results were achieved in 87% of patients.

5. DISCUSSION

Where controversy arises is in differentiating nonobstructive variants and better defining the indications for surgery. Many urinary tract dilations represent distortions of the collecting system that, although at times quite severe, do not represent an obstructive threat to their associated renal moiety. Perinatal ultrasonography has altered the understanding and management of urologic anomalies and dilations, with MGUs, being no exception (3). MGU comprised 20% of antenatally diagnosed urological anomalies. a percentage inordinately urinary tract abnormality, when most were discovered only after they became symptomatic (e.g. infection calculi) and surgery were necessary. (4). Today, through prenatal detection, most children arrive with abnormalities that are totally asymptomatic. If left undetected many MGUs might never become symptomatic, an observation that raises serious questions about treatment. Expectant treatment and serial ultrasonic follow up studies have dramatically redefined the natural history of non-refluxing MCUs.

It is generally agreed that the cause of primary obstructive MGU is an aperistaltic juxta vesical segment 3 to 4 cm long that is unable to propagate urine at acceptable rates of flow. A variety of histologic and ultrastructural abnormalities that alter function have been described. These include disorientation of muscle (5,6), muscular hypoplasia, muscular hypertrophy, and mural fibrosis (7). Excess collagen deposition is a common finding (8,9). Ureteric profilometry shows irregular wave patterns within these segments, so called uretero-arrhythmias(10). The distal ureter is usually involved and may be related to arrested development in the musculature of this segment, which is the last portion of the ureter to develop (5). Regardless of its origin, altered peristalsis prevents the free outflow of urine, and functional obstruction results. Secondary obstructive MGU occurs with neurogenic and non-neurogenic voiding dysfunction or intravesical obstruction such as posterior urethral valves. The dilation that occurs with most of these variants largely resolve once the cause of the elevated intravesical pressures is addressed. In other cases, the ureter remains permanently dilated from what appear to be altered compliance or a permanent insult to the organ’s peristaltic mechanisms or both. Other obstructive causes of ureteral dilation include ureterocoele, bladder diverticula, neurogenic bladder, external compression by retroperitoneal tumour or aberrant vessels. Secondary non-refluxing non-obstructed megaureter can result from acute UTI, accompanied by bacterial endotoxins that inhibit peristalsis. Other causes might be lithium toxicity, diabetes insipidus or mellitus, sickle cell nephropathy and psychogenic polydipsia (11).

Once reflux, obstruction and secondary causes of dilation have been ruled out the designation of primary non-refluxing nonobstructive MGU is appropriate and most new-born MGUs, fall in this category (12,13,14). However, in recent years it has become obvious that at least 50% of cases will have spontaneous resolution. A period of observation is nearly always appropriate when the diagnosis is made in an asymptomatic patient. Because of the high risk of infection 1-2 years of prophylactic antibiotics are recommended in neonates (14). A voiding cystourethrogram is an essential part of the evaluation, not only to rule out reflux but also to ensure that no abnormality of the lower urinary tract is responsible for the upper urinary tract dilation (15). Use of percutaneous renal puncture is occasionally beneficial in the dilated system, it carries minimal risk, making antegrade urography and pressure-flow studies feasible in selected cases. Measurements of the renal pelvic pressure during infusion of saline into the renal pelvis at high rates(10ml/min) (the Whitaker test) may help differentiate nonobstructive from obstructive dilation (18), unfortunately, there is disagreement; clinical judgment is the final arbiter (17). Our patients were treated according to the aetiology, we had two patients with primary refluxing megaureters which were treated by ureteral reimplantation with tapering of the dilated ureter. Figure (1-5).
Fig 1) IVU: Primary Lt. refluxing megaureter
Fig 2) IVU: Primary refluxing megaureter in infant

Fig 3) MCUG: Lt. Refluxing megaureter
Fig 4) IVU: (Postoperative) Megaureter corrected

Fig 5) MCUG (postoperative): Reflux corrected after tapering and reimplantation

Four other cases with primary obstructed megaureter (adynamic segment, stenosis, ureterocoele) were treated with reimplantation and tapering of ureter. One of the two cases of ureterocoele was a complicated case (18-year old girl was diagnosed 10 years before as a case of VUR in one moiety of duplicated ureter and treated with ureteropyeloanastomosis to solve the VUR). Unfortunately, the surgeon didn’t identify the presence of obstructive ureterocoele, and he anastomosed incorrectly the refluxing ureter to the obstructive one, which resulted in complicated urolithiasis in the ureterocoele and ipsilateral kidney. Marsupialization of ureterocoele and extraction of big ureteric calculi and reimplantation with tapering of ureter was performed. Lithotripsy was done later for the kidney stone (Figure 6-8) (18).

Fig 6) KUB: Stone in Rt. Ureterocoele+kidney
Fig 7) IVU: Rt. Ureterocoele with secondary stone

Fig 8) IVU (postoperative) after tapering and reimplantation: Normal urogram

The other case of obstructive ureterocoele in duplicated urinary system with non-functioning dysplastic hydronephrotic upper moiety with severely dilated ureter was treated with marsupialization of ureterocoele and heminephroureterectomy (Figure 9-13) (19).
Among the remaining cases we had 7 cases of secondary megaureters (5 refluxing, 2 obstructive) treated with ablation of posterior urethral valves (Figures 14-15) and optical urethrotomy of stricture urethra.

The dilation resolved once the cause of the elevated intravesical pressure was addressed. Those cases of bladder outlet obstruction (neuropathic bladder) were treated with anticholinergic drugs. The remaining cases, classified as non-refluxing non-obstructed (Figure 16-17) were observed expectantly with serial ultrasonic follow up studies. Good results were achieved in 87% of our cases.

6. CONCLUSIONS

Precise definition of the cause of the dilation and proper patient selection may avoid complications in the reconstructive surgery.

Advancement of understanding of the aetiology and pathophysiology of ureteral dilation may play a role in the differentiation of those megaureters that require surgery to preserve renal function from those that can be observed during its transitional period in the organ's development.

When obstruction is suspected, surgery is indicated.

Patient selection and surgical technique remain the key elements to maintain preservation of renal function.

REFERENCES


