Clinicopathological study of primary obstructive megaureter

C. Saravanan, Karpaga Vinayagam*

Department of Pediatric Surgery, Institute of Child Health and Hospital for Children, Madras Medical College, Egmore, Chennai, Tamil Nadu, India

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*Correspondence:
Dr. Karpaga Vinayagam,
E-mail: arvindr84@gmail.com

ABSTRACT

Background: Megaureter is a non-specific term implying a spectrum of anomalies associated with pathologically excessive ureteral diameter. It implies no particular unifying pathophysiologic principles but merely groups together a spectrum of anomalies associated with increased ureteral diameter. To analyse the clinical profile and various investigation modalities used in diagnosing primary obstructive megaureter.

Methods: This prospective study was conducted in March 2017 to August 2019 at the Institute of Child Health and Hospital for Children (ICH and HC), Egmore, Chennai.

Results: The most common presentation was febrile urinary tract infection, second most presentation was asymptomatic patients, on post-natal evaluation of antenatally diagnosed cases. The most common finding on the antenatal scan is uretero-hydronephrosis. Maximum incidence of ureteric size - between 1 and 1.5 cms. 13 patients underwent surgery after an initial period of observation from the time of presentation. These patients belong to the period of infancy. The coincidence of circular muscle hyperplasia and longitudinal muscle hypoplasia was noted in 18 patients.

Conclusions: In this study, patients presented during infancy were initially observed, prior to surgical intervention. But those patients presented beyond infancy were evaluated and intervened surgically during the same admission. Cohen’s uretero-neocystostomy through intra-vesical approach is the surgical procedure of choice, in the majority of the cases, in this study.

Keywords: Congenital anomalies, Dilated ureter, Primary obstructive megaureter

INTRODUCTION

Megaureter (MGU) is a non-specific term implying a spectrum of anomalies associated with pathologically excessive ureteral diameter.1 It implies no particular unifying pathophysiologic principles but merely groups together a spectrum of anomalies associated with increased ureteral diameter. Because surgical solutions to the anomaly are reliable, the challenge arises in differentiating nonobstructive from obstructive variants and thus better defining the indications for surgery.2 This is analogous to the highly debated management of hydronephrosis caused by UPJ obstruction (Koff and Campbell, 1992), with which MGU may share similar pathophysiologic principles.3 It is apparent by now that all dilatations of the urinary tract do not necessarily translate into physiologically significant obstructive processes with renal functional implications despite the anatomic distortion of the collecting system that they represent.4 Authors knowledge of the megaureter has changed greatly over the past 15 years, primarily due to fetal sonography, which has allowed us to follow the natural history of the megaureter and gain a better insight into treatment. Many cases of antenatally diagnosed MGU will resolve spontaneously.5
METHODS

It is a combined perspective and retrospective study which included patients with primary obstructive megaureter, who attended the pediatric surgery and pediatric urology OPD at the Institute of Child Health and Hospital for Children, Madras Medical College, Chennai. This Prospective study was conducted from March 2017 to August 2019 at the Institute of Child Health and Hospital for Children (ICH and HC) Egmore, Chennai.

Inclusion criteria

All patients with primary obstructive megaureter were proven radiologically and sonographically.

Exclusion criteria

All cases of megaureter with reflux, all cases with bladder dysfunction, urethral obstruction, ureteroceles, and ectopic ureter. The patients were subjected to detailed clinical examination and relevant investigations were performed, namely, ultrasound examination, MCU, IVU, ± DTPA scan. The treatment modalities were studied, and patients were followed up to assess the effectiveness after 6 months of surgery with relevant investigations and extended to the available period. The results were tabulated and analyzed.

RESULTS

Most common presentation was 40% febrile UTI, second most presentation was 20% are asymptomatic patients on post-natal evaluation with antenatal diagnosis. The patients with primary obstructive megaureter are noted predominantly in infants (36.85%) with maximal incidence between 1 to 3 months (6 out of 14 infants). In this study, the sex distribution is found as male: female - 5.3: 1, with definitive male preponderance (Figure 1).

3.26%, which is low in comparison with 10-15% as noted in the study. The percentage of the contralateral renal problem was 7.14%, model, PUJ, distal penile hypospadias, contralateral dysplastic ureter, lower ureteric calculus is most commonly seen (Figure 2).

The most common finding in the antenatal scan is uretero-hydronephrosis. The risk of recurrence or late worsening of hydronephrosis is approximately 1%-5% and applies to all grades of hydronephrosis (Figure 3).

Maximum incidence of ureter size was between 1 and 1.5 cms. In some cases, the ureter may originate from an extrarenal pelvis, a normal anatomic variant in which the renal pelvis lies predominantly outside of the renal sinus. An extrarenal pelvis may be dilated under normal circumstances, but there should be no associated caliectasis. In fact, lack of caliectasis is a key finding differentiating an extrarenal pelvis from a UPJ obstruction at CT urography (Figure 4).

The most common surgical procedure performed aws Cohen's uretero-neocystostomy. One patient underwent Nephroureterectomy at the first instance, due to poorly functioning pyonephrosis. two patients underwent

Figure 1: Symptoms distribution.

The presence of dysplastic or absent contra-lateral kidney in unilateral cases of primary obstructive megaureter, was
surgical procedures prior to definitive ureteric reimplantation. One patient presented at 26 days of life was evaluated. The patient underwent PCN, in view of gross UHN. The other infant presented at 8 months of life, underwent loop ureterostomy in view of the turbid system (Figure 5).

Number of patients who failed to follow up is 5. 19 patients who followed up at six months after surgery were asymptomatic. Febrile UTI was noticed in 13 patients during post-op follow up. Four patients developed VUR during follow up in 32 patients, out of which one patient with grade 3 VUR underwent redo ureteric re-implantation. The other three patients which include one grade 2 VUR and two grade 1 VUR were observed with chemoprophylaxis (Figure 7).

DISCUSSION

Primary obstructive megaureter implies an obstruction to the lower end of a single orthotopic ureter with dilatation of the ureter above the obstruction. It excludes urethral obstruction and vesicoureteric reflux. Primary MGU is a relatively common finding in neonates and infants referred for urologic evaluation. The widespread use of obstetrical ultrasound examination with concomitant fetal screening have changed the age of presentation of congenital uropathies, including megaureter. Currently, about half of the cases are asymptomatic and discovered on prenatal ultrasound. Clinically, patients have UTIs, abdominal pain, fever, pyuria, or hematuria. Microscopic hematuria is frequent and may occur in the absence of infection. This is presumably caused by the disruption of mucosal vessels of the ureter secondary to ureteric distension. Hematuria may also be a sign of calculus formation secondary to urinary stasis. The diagnosis may be made later in life in some asymptomatic patients, either during imaging evaluation for an unrelated complaint or rarely at the time of surgery for another pathologic process. Patients with primary MGU are rarely initially seen with or progress to renal insufficiency. Prenatal ultrasound series suggest UVJ obstruction in up to 23% of patients with urinary tract dilatation. Primary MGU is two to four times more common in boys than girls, has a slight predilection (1.6 to 4.5 times) for the left side, and is bilateral in approximately 25% of patients. The contra-lateral renal problems were noted as MCDK in one patient and PUJ problem in another patient. The presence of dysplastic or absent contra-lateral kidney in unilateral cases of primary obstructive megaureter was 3.26%, which is low in comparison with

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**Figure 4: Distribution of ureter size.**

**Figure 5: Types of surgery.**

Out of 37 patients, only seven patients underwent ureteric remodeling in the form of excisional tapering (18.9%) (Figure 6).

**Figure 6: Ureteric remodelling.**

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10-15% as noted in the study conducted by Hohenfellner et al. The number of the retro-vesical ureter size found in the range between more than 1 and 1.5 were 21. There was a significant improvement in the renal function by DTPA scan from 20% at the time of loop ureterostomy-left side (8 months of life) to 40% at the time of ureteric reimplantation (24 months of life). During the definitive surgical procedure, Ureteric remodeling was performed in only 7 out of 37 patients their improvement was compared with the study conducted by Sanyanusin et al.

CONCLUSION

In this study, the most common symptom at presentation is febrile UTI (63.15%), followed by asymptomatic patients (26.32%) The contra-lateral renal dysplasia in unilateral cases is found to be 3.26%. The majority of cases with retro-vesical ureter size lie between 1 and 1.5 cm (48.8%), which are not considered to belong to the spontaneously resolving group. MCU and IVU are the most common investigational modality performed. In this study, patients presented during infancy were initially observed prior to surgical intervention. But those patients presented beyond infancy were evaluated and intervened surgically during the same admission. Cohen’s uretero-neocystostomy through intra-vesical approach is the surgical procedure of choice, in majority of the cases, in this study. The re-implantation failure was noted in 28.12% (includes persistent obstruction and minimal grades of VUR).

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REFERENCES
