Journal of Pediatric Urology (2014) 10, 26—33

Journal of Pediatric urology

REVIEW ARTICLE

British Association of Paediatric Urologists consensus statement on the management of the primary obstructive megaureter

Marie-Klaire Farrugia [a,*], Rowena Hitchcock [b], Anna Radford [c], Tariq Burki [d], Andrew Robb [e], Feilim Murphy [f], On behalf of the British Association of Paediatric Urologists (BAPU)

[a] *Department of Paediatric Surgery and Urology, Chelsea and Westminster Hospital, London, UK*
[b] *Department of Paediatric Surgery and Urology, John Radcliffe Hospital, Oxford, UK*
[c] *Department of Paediatric Surgery and Urology, Leeds General Infirmary, Leeds, UK*
[d] *Department of Paediatric Urology, Southampton University Hospital Trust, Southampton, UK*
[e] *Department of Paediatric Urology, Birmingham Children's Hospital, Birmingham, UK*
[f] *Department of Paediatric Surgery and Urology, St George's Hospital, London, UK*

Received 21 April 2013; accepted 16 September 2013
Available online 16 October 2013

KEYWORDS
Primary obstructive megaureter;
Ureteral obstruction;
Ureteral reimplantation;
Ureteral stent;
Antenatal hydronephrosis;
Consensus statement

Abstract *Introduction:* It is well-known that the majority of congenital megaureters may be managed conservatively, but the indications and surgical options in patients requiring intervention are less well defined. Hence this topic was selected for discussion at the 2012 consensus meeting of the British Association of Paediatric Urologists (BAPU). Our aim was to establish current UK practice and derive a consensus management strategy.
Methods: An evidence-based literature review on a predefined set of questions on the management of the primary congenital megaureter was presented to a panel of 56 Consultant Surgeon members of the British Association of Paediatric Urologists (BAPU), and current opinion and practice established. Each question was discussed, and a show of hands determined whether the panel reached a consensus (two-thirds majority).
Results: The BAPU defined a ureteric diameter over 7 mm as abnormal. The recommendation was for newborns with prenatally diagnosed hydroureteronephrosis to receive antibiotic prophylaxis and be investigated with an ultrasound scan and micturating cystourethrogram, followed by a diuretic renogram once VUR and bladder outlet obstruction had been excluded. Initial management of primary megaureters is conservative. Indications for surgical

* Corresponding author. Chelsea and Westminster Hospital, 369 Fulham Road, London SW10 9NH, UK.
E-mail address: Marie-Klaire.Farrugia@Chelwest.nhs.uk (M.-K. Farrugia).

http://dx.doi.org/10.1016/j.jpurol.2013.09.018

intervention include symptoms such as febrile UTIs or pain, and in the asymptomatic patient, a DRF below 40% associated with massive or progressive hydronephrosis, or a drop in differential function on serial renograms. The BAPU recommended a ureteral reimplantation in patients over 1 year of age but recognized that the procedure may be challenging in infancy. Proposed alternatives were the insertion of a temporary JJ stent or a refluxing reimplantation.
Conclusion: A peer-reviewed consensus guideline for the management of the primary megaureter has been established. The guideline is based on current evidence and peer practice and the BAPU recognized that new techniques requiring further studies may have a role in future management.

Introduction

Congenital anomalies of the vesicoureteral junction (VUJ) often present because of detection of an associated dilated or 'mega'-ureter. Many synonyms for this condition exist, including hydroureter and megaloureter, but all represent a ureter with a diameter larger than normal with or without associated renal pelvis dilatation. Smith [1] classified megaureters into four categories, obstructed, refluxing, refluxing with obstruction, and non-refluxing/non-obstructing, later subdivided into primary and secondary by King [2]. The Pfister—Hendren classification established in 1978 was based on the morphological appearance: type I involved the distal ureter without associated hydronephrosis; type II extended to both ureter and pelvis; and type III was associated with severe hydroureteronephrosis and ureteric tortuosity [3,4]. It is well known that the majority of megaureters may be managed conservatively, but the indications and surgical options in patients requiring intervention are less well defined. Hence this topic was selected for discussion at the 2012 consensus meeting of the British Association of Paediatric Urologists (BAPU).

The BAPU (www.bapu.org.uk) was founded in Cambridge in 1992 by two eminent paediatric urologists, Philip Ransley and Robert Whitaker. The association represents the views of the membership of paediatric urologists on matters of standards and ethics, and supports practice, research, and training in paediatric urology. The consensus section to our annual meeting was established in 2011 with the aim of establishing peer practice within BAPU and ensuring that our practice is in line with established studies and guidelines. The 2012 meeting focused on the management of the primary obstructive megaureter.

Methods

Four paediatric urology fellows were asked to search the literature for evidence relating to the definition, pathophysiology, prenatal findings, postnatal investigation, initial management, and surgical options for the primary obstructive megaureter (POM). Using the NCBI PubMed and Medline databases, articles were sought up to the 1 July 2012. The terms megaureter, megaloureter, ureterovesical junction, vesicoureteric junction obstruction, obstructive congenital hydronephrosis, foetal ureteral obstruction, hydroureter, and large ureter were used; Boolean operators were utilized. A total of 2205 articles were identified; non-English language publications were excluded, leaving 1531 reports to review. These were then divided further for specific aspects of the review: definition, natural history, pathology, classification, diagnosis, imaging, and treatment. Papers relating to management of megaureter were analysed further for use of antibiotics, ureteral reimplantation, stents, balloon dilatation, endoureterotomy, ureterostomy, and follow-up. Additional referenced information has been taken from reports published after July 2012, guidelines, and a published textbook.

The evidence was presented to 56 consultant paediatric urologists present at the 2012 annual BAPU meeting. Specific questions were posed by the chair (FM) and a show of hands used to determine whether a consensus (two-thirds majority) was reached. All questions raised, regardless of consensus, are included in this study. In order to streamline the discussion, BAPU agreed that the focus of the consensus would be the patient unresponsive to conservative management, in particular when intervention is required below 1 year of age. The refluxing megaureter was excluded from the discussion.

Definition and pathophysiology

The definition of a dilated ureter is not well established in the literature. Cussen [5] examined ureters of foetuses over 20 weeks' gestation and children up to the age of 12 years and established mean ureteral dimensions for foetuses at 30 weeks' gestation, and for infants and children at 3 months and 3, 6, and 12 years. The upper limit of the range of diameter of the mid-section of the ureter from birth to 12 years was 0.5—0.65 cm; therefore, the upper limit of normal was regarded as closely approximating 0.7 cm. Hellstrom et al. [6] provided radiological data of normal ureteric diameter in 194 children aged 0—16 years. This paper has resulted in the radiological definition of a dilated ureter as that above 7 mm in diameter. On the basis of these studies, the BAPU was asked to determine the diameter at which a ureter would be considered to be "dilated".

Consensus: Retrovesical ureteric diameter $\geq$ 7 mm from 30 weeks' gestation onwards is abnormal.

A number of studies in the literature have looked into the possible pathogenesis of a congenital megaureter. The timing of smooth muscle differentiation in the distal ureter is unknown, but may be key to understanding why reflux or obstruction develop in utero, persist in the newborn, and often subside later in childhood. Pirker et al. [7] studied

bladder specimens form porcine foetuses and piglets. They found that the intravesical ureter did not mature until after birth. The time-course of muscular development in this study correlates with findings in humans, in whom smooth muscle bundles of the extramural ureter first appear in weeks 17—22 of gestation, and ureteral muscle development proceeds in a craniocaudal direction [8]. Tanagho [9] also observed that the distal ureter is the last portion to develop its muscular coat, and that early muscular differentiation is primarily of the circular muscles. The process whereby the circular muscle pattern, which is typical of the foetal ureter, changes progressively into the double muscle layers of the full-term infant, may last up to 2 years, and may explain the transient functional disturbances of the VUJ during this period.

Prenatal diagnosis

In most UK centres, prenatal anomaly scans are performed at 18—20 weeks' gestation. Data form a large European database for surveillance of congenital malformations (EUROCAT) collected over a 10-year period gave an overall prevalence of congenital hydronephrosis of 11.5 per 10,000 births; 73% of cases were prenatally diagnosed, and 72% were male [10]. The incidence of prenatally-diagnosed megaureter is not known. This may be because detection of a dilated ureter in utero by ultrasound is a difficult endeavour [11]. The recent Society for Foetal Urology consensus statement could also find no evidence correlating the prenatal finding of a dilated ureter with postnatal outcome [12]. Based on the definition of a dilated ureter on prenatal ultrasound, the BAPU was asked whether they would investigate a dilated ureter postnatally (with or without associated hydronephrosis).

Consensus: A prenatal ureteral diameter over 7 mm is abnormal and should be investigated postnatally.

Postnatal investigation

Newborns with a prenatal diagnosis of hydronephrosis are usually started on antibiotic prophylaxis, until a definitive diagnosis is made. Song et al. [13] showed that the risk of UTI is higher with VUJ than pelviureteric junction (PUJ) obstruction, and that UTIs in this group tend to occur within the first 6 months of life. A long-term outcome study of POMs suggested a surprisingly high UTI rate in these patients, with 35% of patients requiring at least one hospital admission [14]. The incidence of UTIs in untreated infants with POM was found to be 0.94 per year. Antibiotic prophylaxis reduced this incidence by 83% in the first 6 months and 55% in the first year of life.

Consensus: In the presence of hydroureteronephrosis, antibiotic prophylaxis is advisable for the first 6—12 months of life.

The BAPU consensus was that the initial postnatal investigation in babies with unilateral ureteric dilatation should be a renal tract ultrasound scan. Boys with bilateral ureteric dilatation or hydroureteronephrosis should be scanned sooner and an early micturating cystourethrogram (MCUG) obtained in order to exclude bladder outlet obstruction (BOO) such as posterior urethral valves (PUVs). The BAPU raised the issue that boys with unilateral hydroureteronephrosis (with or without an abnormal bladder) should also have an early MCUG to exclude PUV, which may be associated with unilateral dilatation on ultrasound scan in up to 14% of PUV patients [15].

Consensus: All babies with prenatally diagnosed ureteric dilatation should have a postnatal ultrasound scan. Babies with bilateral ureteric dilatation and boys with unilateral hydroureteronephrosis should have an early MCUG to exclude bladder outlet obstruction. An MCUG is indicated in all patients to exclude the presence of vesicoureteral reflux (VUR).

In the absence of urethral obstruction or VUR, a ^{99m}Tc-MAG-3 diuretic renogram (MAG-3) is indicated to look for evidence of obstruction at the VUJ. The BAPU consensus was that this study is indicated in the presence of hydroureteronephrosis. When asked whether a MAG-3 was indicated in the presence of *isolated* ureteric dilatation, the BAPU members said they would only investigate isolated ureteric dilatation $>$ 10 mm, based on data from the Great Ormond Street Hospital series [16].

Consensus: Once BOO and VUR are excluded, a MAG-3 scan is indicated in babies with hydroureteronephrosis or isolated ureteric dilatation $>$ 10 mm to look for obstruction at the VUJ.

Babies presenting postnatally (e.g. with a urinary tract infection) are also investigated in a similar fashion, although it is wise to wait for recovery from urosepsis before embarking on an MCUG or MAG-3 [11].

Interpretation of the MAG-3 renogram in the presence of a dilated ureter may be difficult, as delayed transit may be caused by the increased capacity afforded by a dilated pelvis and ureter per se [17]. Poor drainage may also be apparent because the bladder is full, or because the effect of gravity on drainage is incomplete. For all these reasons, interpretation of the washout curve should be made in the light of the differential renal function (DRF) and the degree of renal pelvis dilatation. Normal values of DRF are between 45% and 55% uptake [18]. DRF should be interpreted in clinical context, since values within the normal range may be seen also when there is bilateral renal damage and/or in the presence of chronic renal failure. The BAPU panel considered an initial DRF $<$ 40%, or a drop in DRF by 5% on serial scans, to be significant. On the other hand, delayed transit on MAG-3 in the presence of stable or improving dilatation, and a DRF above 40%, in an asymptomatic patient, are not strong indicators of obstruction, although continued follow-up would be indicated.

Consensus: A combination of clinical and radiological findings must be taken into account when diagnosing a megaureter as "obstructed" or "non-obstructed". In the asymptomatic patient, the BAPU considered the presence of an initial DRF below 40%, or a drop in DRF of 5% on serial scans, and/or increasing dilatation on serial ultrasound scans, to be suggestive of obstruction. Delayed transit on MAG-3 in the presence of stable or improving dilatation, and a DRF above 40%, in an asymptomatic patient, were not felt to be strong indicators of obstruction.

Management

The management of primary megaureters has evolved over the past 20 years. In 1989, Peters et al. [19] reported that

89% of patients required surgery before 8 months of age. That same year, Keating et al. [20] reported that when the decision to intervene was based on absolute renal function, up to 87% of patients could be followed up conservatively. A further publication from the same unit confirmed stability or improvement at long-term follow-up of the same group of patients who were managed conservatively [21]. The initial conservative management of megaureters has since been well established, with a number of outcome studies confirming this. A recent long-term follow-up study of 74 POMs (distal ureter 7 mm and above) initially managed conservatively revealed that at a median 10-year follow-up, 73% did not require surgical intervention. On univariate analysis, age at presentation (prenatal vs. postnatal), megaureter type, differential renal function (<40%), and washout pattern were significant predictors of spontaneous resolution. However, on multivariate analysis, only age at presentation and washout pattern were significant predictors. Surgical indications included urosepsis or massive dilatation with decreased renal function at presentation, or onset of symptoms or obstruction with loss of function at follow-up. Long-term follow-up was recommended as symptoms could develop after years of observation [22].

The BAPU supported initial conservative management. When asked what, in their practice, were the indicators for surgical intervention, key indicators were felt to be an initial DRF < 40%, especially when associated with massive hydroureteronephrosis, and failure of conservative management (breakthrough febrile UTIs, pain, worsening dilatation, or deteriorating DRF).

The panel was then questioned on choice of surgical intervention. In babies over 1 year of age, the procedure of choice for the majority was a ureteric reimplantation with or without ureteral tapering. However, the panel agreed that the reimplantation of a grossly dilated ureter into a small infantile bladder could be a challenging operation in babies below 1 year of age, and a number of alternative temporizing or definitive options were then put forward for discussion. These included:

- temporary double-J stenting
- endoscopic balloon dilatation
- endoureterotomy
- cutaneous ureterostomy
- refluxing ureteral reimplantation.

The BAPU panel was also asked what emergency intervention they would perform when faced with a septic child with an obstructed infected system not responsive to intravenous antibiotics, and the majority voted in favour of a percutaneous nephrostomy or cutaneous ureterostomy in preference to a double-J stent.

Ureteric reimplantation

Peters et al. [19] showed that repair of an obstructed megaureter in early infancy improves renal drainage as assessed by excretory urography, and therefore offers the potential for preventing renal damage before the development of symptoms or decline in function on the renogram. Ureteric reimplantation allows the aperistaltic, narrow segment to be excised, and the ureter to be tunnelled into the bladder in an antireflux fashion. According to a study by Paquin [23] in 1959, the tunnel should be at least five times the ureteral diameter to be effective, a ratio that may be used as a guide for effective reimplantation. In order to obtain this ratio when the ureteral diameter is above 10 mm, the distal 10 cm of the ureter may need to be tapered. A psoas hitch may also be required in order to ensure a straight entry of the ureter into the bladder. The two popular techniques used for tailoring megaureters are plication as reported by Starr [24] and Kalicinski et al. [25], and tapering as modified by Hendren [26]. Kalicinski et al. [25] described a folding technique whereby the lateral, avascular part of the ureter was excluded from the lumen by a longitudinal running suture, and then folded posteriorly prior to reimplantation. Starr [24] modified this procedure, introducing many interrupted Lembert sutures that folded the ureteral wall inwards, thus preserving its blood supply but making the subsequent reimplantation more cumbersome. Hendren [26] advised excisional tapering, whereby a longitudinal strip of the redundant ureter is excised, and the ureter sutured in two layers. Ben-Meir et al. [27] compared the outcome of megaureter reimplantation with and without tailoring, and showed that there was no significant difference in outcome (94% success rate in the tailored group, and 96% in the non-tailored group). Intravesical reimplantation (Cohen, and Politano—Leadbetter) techniques are preferable when the ureter has been tailored, especially in older children with a degree of bladder dysfunction [28]. Laparoscopic and robotic ureteric tapering and implantation techniques have also been described [29,30].

Ureteric reimplantation in infants below 1 year of age may be challenging due to the discrepancy between the grossly dilated ureter and the small infantile bladder, and concern regarding possible iatrogenic bladder dysfunction. de Jong [31] investigated the bladder functional outcome of 49 neonates and infants with primary obstructive or refluxing megaureters who had undergone a Politano—Leadbetter procedure, at a mean follow-up of 7.3 years. No onset of voiding dysfunction was noted at follow-up, and bladder capacity for age was significantly increased [31]. No postoperative ureteral obstruction was observed. The study concluded that early major reconstructions of the lower urinary tract do not damage the urodynamic properties of the bladder and pelvic floor, provided that the surgery is performed by specialized paediatric urological surgeons. A further study from the same group [32] compared urodynamic outcome in patients who had undergone ureteral reimplantation before or after 1 year of age. No significant difference in bladder capacity, compliance, or incidence of instability was found. Upadhyay et al. [33] showed equally encouraging results at a 9.5-year follow-up study of 21 infants who had undergone ureteral reimplantation (half the patients had bilateral procedures) before 12 months of age. Nerve-sparing techniques are also being implemented in laparoscopic and robotic reimplantation techniques, with promising outcomes in terms of bladder function [34]. The panel therefore concluded that evidence for the commonly held belief that early reimplantation carried a risk of bladder dysfunction was weak.

Consensus: The BAPU panel agreed that ureteric reimplantation below 1 year of age is feasible, but the majority of the group felt that they would consider alternative

temporizing or permanent interventions and avoid reimplantation in infancy. There was no strong consensus regarding reimplantation technique or tapering versus non-tapering.

Endoscopic procedures

In 1999, Shenoy and Rance [35] reported of the use of JJ stents as a temporizing measure in infants with POMS. The procedure involves the insertion of a 4.7F or 5.2F double pigtail stent endoscopically where possible. A tight VUJ obstruction may preclude endoscopic insertion, in which case an open insertion via a cystostomy combined with dilatation of the ureteric orifice using a lachrymal probe may be performed. More recently, two UK units have reported their long-term outcomes. Farrugia et al. [36] reviewed the Southampton experience in utilizing stenting as a temporizing procedure in infants less than 1 year of age. Stents were left in situ for 6 months and then replaced if the baby was still below 1 year of age. Complications (stent migration, stone formation, or infection) occurred in 31%. At follow-up after stent removal, drainage improved in 56% of cases, in whom no further intervention was required [36]. Carroll et al. [37] reported the Birmingham experience with the use of JJ stents in patients of all ages. Thirty-one patients age 2 months to 18 years were stented over a 7-year period. One patient sustained an iatrogenic ureteric injury requiring immediate reimplantation. Of the remainder, 15 (50%) had recurrent hydroureteronephrosis after the initial stent was removed and eight patients were reimplanted because of functional impairment. Seven patients were restented, of which only two patients subsequently required reimplantation, giving a 67% overall success rate [36].

Angerri et al. [38] published their experience with endoscopic balloon dilatation in a series of seven patients aged 1—3 years, using a 4F dilating balloon insufflated to 12—14 atm for 3—5 min. Disappearance of the narrowed ring was verified radiologically. A JJ stent was left in situ for 2 months. Postoperative drainage improved in five out of seven patients after one or two dilatations at short-term follow-up. Garcia-Aparicio et al. [39] published their experience of high-pressure balloon dilatation in babies with a median age of 7 months. The VUJ was dilated at 14 atm for 3 min under direct fluoroscopic control until disappearance of the narrow ring was observed. A double-J stent was then left in situ for 8 weeks. Of 13 patients, five required a further dilatation and three patients required reimplantation at a median follow-up of 25 months.

Kajbafzadeh et al. [40] described an endoureterotomy procedure in patients with "intravesical ureteral obstruction" and a stricture length < 1.5 cm. Their case series of 47 patients includes infants as young as 1.5 months, although the mean age at intervention was 3.7 years. The authors report an impressive success rate of 90% at a mean follow-up of 39 months, including 71% complete resolution of hydronephrosis, with self-limiting haematuria as the only complication.

Cutaneous ureterostomy

A temporary cutaneous ureterostomy proximal to the obstruction is an alternative option, allowing good decompression of the obstructed system and improvement in ureteric dilatation, thus facilitating definitive reimplantation. However, the procedure may be complicated by an 8—22% incidence of stomal stenosis [41—43] and pyelonephritis in up to 31% of patients, despite the use of prophylactic antibiotics [44,45]. In bilateral cases, cutaneous ureterostomies will result in bladder defunctionalization and the potential long-term loss of bladder capacity. It is also possible that ureterostomies may prove difficult to manage by the parents.

Refluxing ureteral reimplantation

The refluxing ureteral reimplant was first described by Lee et al. [46] as a temporizing solution to the obstructed megaureter requiring intervention in infancy. The authors described a procedure whereby the ureter was transected proximal to the narrow segment and anastomosed to the lateral aspect of the bladder in a freely refluxing fashion, thus converting "dangerous" obstruction to the lesser evil that is reflux. More recently, the technique has been simplified by leaving the obstructed segment in situ and performing a side-to-side ureteroneocystotomy proximal to the obstruction (M. Koyle, Hospital for Sick Children, Toronto, presented at BAPU Consensus meeting). The authors proposed this procedure as an option to cutaneous ureterostomy, with the added advantage of maintaining bladder cycling. Kaefer et al. [47] recently presented the outcome of 13 patients (16 obstructed ureters) who had undergone internal diversion at a mean age of 2.5 months. All patients demonstrated improved drainage of the affected kidney following surgery. Postoperative urinary tract infection occurred in two patients. Definitive surgical treatment was undertaken in 14 out of 16 ureters (ureteral reimplantation with [9] or without [3] tapering, and nephrectomy [2]). The first four patients were stented, but subsequent patients were managed without stents. The authors questioned whether a secondary definitive procedure to reimplant the ureter is of absolute necessity. In their view, the presence of a large reservoir of urine that may never completely empty with voiding, possesses significant risk of future infection once prophylaxis is discontinued, and hence a definitive procedure advisable.

Consensus: Temporizing JJ stents and cutaneous ureterostomies were the most common currently performed procedures by the BAPU surgeons in babies below 1 year of age and were felt to be safe and acceptable options. There was very limited experience with endoscopic balloon dilatation. Although no outcome studies are as yet available, the refluxing ureteral reimplantation gained BAPU support and the majority of the group stated they would consider performing this procedure in preference to a ureterostomy in future. Where surgical intervention is indicated in infancy, BAPU proposed the option of endoscopic stenting as first line, followed by a refluxing reimplantation if endoscopic insertion failed.

Follow-up

Long-term follow-up of conservatively managed megaureters is warranted due to evidence of late recurrence in teenage and adult patients. A long-term follow-up study by Shukla et al. [48] reported late deterioration in a 14-year-

Table 1 British Association of Paediatric Urologists (BAPU) consensus summary.

Definition of a megaureter	Retrovesical ureteric diameter $\geq$ 7 mm from 30 weeks' gestation onwards.
Postnatal management	Newborns with prenatal hydroureteronephrosis should be started on antibiotic prophylaxis; in conservatively managed cases, prophylaxis is recommended for the first 6—12 months of life.
Investigation	All babies with prenatal ureteric dilatation should have a postnatal ultrasound scan. Babies with bilateral ureteric dilatation and boys with unilateral hydroureteronephrosis should have an *early* MCUG to exclude bladder outlet obstruction. An MCUG is indicated in all patients to exclude the presence of VUR. Once BOO and VUR are excluded, a MAG-3 scan is indicated in babies with hydroureteronephrosis or isolated ureteric dilatation $>$ 10 mm to look for obstruction at the VUJ.
Defining "obstruction"	A combination of clinical and radiological findings must be taken into account when diagnosing a megaureter as "obstructed". In the asymptomatic patient, the BAPU considered the presence of an initial DRF below 40%, or a drop in DRF of 5% on serial scans, and/or increasing dilatation on serial ultrasound scans, to be suggestive of obstruction. Delayed transit on MAG-3 in the presence of stable or improving dilatation, and a DRF above 40%, in an asymptomatic patient, were not felt to be strong indicators of obstruction.
Initial management	The BAPU recommended initial conservative management. Indications for surgical intervention include an initial DRF $<$ 40% especially when associated with massive hydroureteronephrosis, and failure of conservative management (breakthrough febrile UTIs, pain, worsening dilatation or deteriorating DRF on serial scans).
Surgical intervention	The BAPU panel agreed that although ureteric reimplantation below one year of age is feasible and safe, the majority of the group felt that they would consider alternative interventions in infants. The most favoured strategy was of temporary endoscopic stenting as first line, followed by a refluxing reimplantation if endoscopic insertion failed.
Follow-up	Long-term follow-up is warranted for conservatively managed megaureters as symptoms could occur later in childhood or even in adulthood.

BOO = bladder outlet obstruction; DRF = differential renal function; MCUG = micturating cystourethrogram; UTI = urinary tract infection; VUR = vesicoureteral reflux; VUJ = vesicoureteral junction.

old boy with bilateral primary megaureters who presented with increased dilatation and functional deterioration 6 years after he was discharged from conservative follow-up. More alarming is the study by Hemal et al. [49], who reported the outcome of 55 patients with congenital megaureters presenting for the first time in the third decade of life, of whom 20 patients were found to have renal calculi. Five patients with bilateral megaureters were in chronic renal failure at diagnosis, of whom two eventually died of the disease.

In the UK, paediatric urologists generally follow up patients up to the age of 16 or 18 years and would then transition to an adult service. There was no consensus on how long follow-up was advisable.

Consensus: Long-term follow-up throughout childhood is warranted for conservatively managed megaureters.

Conclusion

A summary of the BAPU Consensus statement is presented in Table 1. Having considered the available peer-reviewed evidence on the management of POM, the BAPU defined a ureteric diameter over 7 mm as abnormal. The recommendation was for newborns with prenatally diagnosed hydroureteronephrosis to receive antibiotic prophylaxis and be investigated with an ultrasound scan and MCUG, followed by a diuretic renogram once VUR and bladder outlet obstruction have been excluded. Initial management of primary megaureters is conservative. Indications for surgical intervention include symptoms such as febrile UTIs or pain, and, in the asymptomatic patient, a DRF below 40% associated with massive or progressive hydronephrosis, or a drop in differential function on serial renograms. The BAPU recommended a ureteral reimplantation in patients over 1 year of age but recognized that the procedure may be challenging in infancy. Proposed alternatives were the insertion of a temporary JJ stent or a refluxing reimplantation.

Conflict of interest

None.

Funding

None.

Ethical approval

No ethical consent required.

Acknowledgements

The authors would like to thank members of the British Association of Paediatric Urologists, and invited guests, Professors John Hutson and Martin Koyle, for their contribution to the consensus discussion and statement. We

thank Professor Martin Kaefer for his contribution and data regarding the refluxing ureteral reimplantation.

References

[1] Smith ED. Report of Working Party to establish the international nomenclature for the large ureter. In: Bergsma D, Duckett J, editors. Birth defects. Original articles series; 1977. p. 3—8.

[2] King LR. Megaloureter: definition, diagnosis and management. J Urol 1980;123:222—3.

[3] Pfister RC, Hendren WH. Primary megaureter in children and adults. Clinical and pathophysiologic features of 150 ureters. Urology 1978;12:160—76.

[4] Weber AL, Pfister RC, James Jr AE, Hendren WH. Megaureter in infants and children: roentgenologic, clinical, and surgical aspects. Am J Roentgenol Radium Ther Nucl Med 1971;112:170—7.

[5] Cussen LJ. Dimensions of the normal ureter in infancy and childhood. Invest Urol 1967;5:164—78.

[6] Hellstrom M, Hjalmas K, Jacobsson B, Jodal U, Oden A. Normal ureteral diameter in infancy and childhood. Acta Radiol Diagn (Stockh) 1985;26:433—9.

[7] Pirker ME, Rolle U, Shinkai T, Shinkai M, Puri P. Prenatal and postnatal neuromuscular development of the ureterovesical junction. J Urol 2007;177:1546—51.

[8] Matsuno T, Tokunaka S, Koyanagi T. Muscular development in the urinary tract. J Urol 1984;132:148—52.

[9] Tanagho EA. Intrauterine fetal ureteral obstruction. J Urol 1973;109:196—203.

[10] Garne E, Loane M, Wellesley D, Barisic I. Congenital hydronephrosis: prenatal diagnosis and epidemiology in Europe. J Pediatr Urol 2009;5:47—52.

[11] Farrugia M. Ureterovesical junction anomalies. In: Wilcox D, Godbole C, Cooper C, editors. Pediatric urology book; 2012.

[12] Nguyen HT, Herndon CD, Cooper C, Gatti J, Kirsch A, Kokorowski P, et al. The Society for Fetal Urology consensus statement on the evaluation and management of antenatal hydronephrosis. J Pediatr Urol 2010;6:212—31.

[13] Song SH, Lee SB, Park YS, Kim KS. Is antibiotic prophylaxis necessary in infants with obstructive hydronephrosis? J Urol 2007;177:1098—101.

[14] Gimpel C, Masioniene L, Djakovic N, Schenk JP, Haberkorn U, Tonshoff B, et al. Complications and long-term outcome of primary obstructive megaureter in childhood. Pediatr Nephrol 2010;25:1679—86.

[15] Williams CR, Perez LM, Joseph DB. Accuracy of renal-bladder ultrasonography as a screening method to suggest posterior urethral valves. J Urol 2001;165(Pt 2):2245—7.

[16] Liu HY, Dhillon HK, Yeung CK, Diamond DA, Duffy PG, Ransley PG. Clinical outcome and management of prenatally diagnosed primary megaureters. J Urol 1994;152(Pt 2):614—7.

[17] Gordon I. Diuretic renography in infants with prenatal unilateral hydronephrosis: an explanation for the controversy about poor drainage. BJU Int 2001;87:551—5.

[18] Gordon I, Piepsz A, Sixt R. Guidelines for standard and diuretic renogram in children. Eur J Nucl Med Mol Imaging 2011;38:1175—88.

[19] Peters CA, Mandell J, Lebowitz RL, Colodny AH, Bauer SB, Hendren WH, et al. Congenital obstructed megaureters in early infancy: diagnosis and treatment. J Urol 1989;142(Pt 2):641—5.

[20] Keating MA, Escala J, Snyder III HM, Heyman S, Duckett JW. Changing concepts in management of primary obstructive megaureter. J Urol 1989;142(Pt 2):636—40.

[21] Baskin LS, Zderic SA, Snyder HM, Duckett JW. Primary dilated megaureter: long-term followup. J Urol 1994;152(Pt 2):618—21.

[22] Di RD, Aguiar L, Cascini V, Di NM, McCarten KM, Ellsworth PI, et al. Long-term followup of primary nonrefluxing megaureter. J Urol 2013.

[23] Paquin Jr AJ. Ureterovesical anastomosis: the description and evaluation of a technique. J Urol 1959;82:573—83.

[24] Starr A. Ureteral plication. A new concept in ureteral tailoring for megaureter. Invest Urol 1979;17:153—8.

[25] Kalicinski ZH, Kansy J, Kotarbinska B, Joszt W. Surgery of megaureters — modification of Hendren's operation. J Pediatr Surg 1977;12:183—8.

[26] Hendren WH. Operative repair of megaureter in children. J Urol 1969;101:491—507.

[27] Ben-Meir D, McMullin N, Kimber C, Gibikote S, Kongola K, Hutson JM. Reimplantation of obstructive megaureters with and without tailoring. J Pediatr Urol 2006;2:178—81.

[28] DeFoor W, Minevich E, Reddy P, Polsky E, McGregor A, Wacksman J, et al. Results of tapered ureteral reimplantation for primary megaureter: extravesical versus intravesical approach. J Urol 2004;172(Pt 2):1640—3.

[29] Ansari MS, Mandhani A, Khurana N, Kumar A. Laparoscopic ureteral reimplantation with extracorporeal tailoring for megaureter: a simple technical nuance. J Urol 2006;176(Pt 1):2640—2.

[30] Hemal AK, Nayyar R, Rao R. Robotic repair of primary symptomatic obstructive megaureter with intracorporeal or extracorporeal ureteric tapering and ureteroneocystostomy. J Endourol 2009;23:2041—6.

[31] de Jong TP. Treatment of the neonatal and infant megaureter in reflux, obstruction and complex congenital anomalies. Acta Urol Belg 1997;65:45—7.

[32] de Kort LM, Klijn AJ, Uiterwaal CS, De Jong TP. Ureteral reimplantation in infants and children: effect on bladder function. J Urol 2002;167:285—7.

[33] Upadhyay J, Shekarriz B, Fleming P, Gonzalez R, Barthold JS. Ureteral reimplantation in infancy: evaluation of long-term voiding function. J Urol 1999;162(Pt 2):1209—12.

[34] Casale P, Patel RP, Kolon TF. Nerve sparing robotic extravesical ureteral reimplantation. J Urol 2008;179:1987—9.

[35] Shenoy MU, Rance CH. Is there a place for the insertion of a JJ stent as a temporizing procedure for symptomatic partial congenital vesico-ureteric junction obstruction in infancy? BJU Int 1999;84:524—5.

[36] Farrugia MK, Steinbrecher HA, Malone PS. The utilization of stents in the management of primary obstructive megaureters requiring intervention before 1 year of age. J Pediatr Urol 2011;7:198—202.

[37] Carroll D, Chandran H, Joshi A, McCarthy LS, Parashar K. Endoscopic placement of double-J ureteric stents in children as a treatment for primary obstructive megaureter. Urol Ann 2010;2:114—8.

[38] Angerri O, Caffaratti J, Garat JM, Villavicencio H. Primary obstructive megaureter: initial experience with endoscopic dilatation. J Endourol 2007;21:999—1004.

[39] Garcia-Aparicio L, Rodo J, Krauel L, Palazon P, Martin O, Ribo JM. High pressure balloon dilation of the ureterovesical junction — first line approach to treat primary obstructive megaureter? J Urol 2012;187:1834—8.

[40] Kajbafzadeh AM, Payabvash S, Salmasi AH, Arshadi H, Hashemi SM, Arabian S, et al. Endoureterotomy for treatment of primary obstructive megaureter in children. J Endourol 2007;21:743—9.

[41] Kogan BA, Gohary MA. Cutaneous ureterostomy as a permanent external urinary diversion in children. J Urol 1984;132:729—31.

[42] MacGregor PS, Kay R, Straffon RA. Cutaneous ureterostomy in children — long-term followup. J Urol 1985;134:518—20.

[43] Sarduy GS, Crooks KK, Smith JP, Wise HA. Results in children managed by cutaneous ureterostomy. Urology 1982;19:486—8.

[44] Gearhart JP. Primary obstructive ureter in neonates. Treatment by temporary uretero-cutaneostomy. Br J Urol 1994;74:133—4.
[45] Kitchens DM, DeFoor W, Minevich E, Reddy P, Polsky E, McGregor A, et al. End cutaneous ureterostomy for the management of severe hydronephrosis. J Urol 2007;177:1501—4.
[46] Lee SD, Akbal C, Kaefer M. Refluxing ureteral reimplant as temporary treatment of obstructive megaureter in neonate and infant. J Urol 2005;173:1357—60.
[47] Kaefer M, Misseri R, Gitlin J, Frank E, Rhee A, Balanos J, et al. Refluxing ureteral reimplantation: a logical method for managing neonatal UVJ obstruction. In: Presented at the 2012 European Society for Pediatric Urology (ESPU) annual meeting; 9—12 May 2012.
[48] Shukla AR, Cooper J, Patel RP, Carr MC, Canning DA, Zderic SA, et al. Prenatally detected primary megaureter: a role for extended followup. J Urol 2005;173:1353—6.
[49] Hemal AK, Ansari MS, Doddamani D, Gupta NP. Symptomatic and complicated adult and adolescent primary obstructive megaureter — indications for surgery: analysis, outcome, and follow-up. Urology 2003;61:703—7.