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REVIEW ARTICLE

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

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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

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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