MEGA-URETER

By D. INNES WILLIAMS, M.D., M.CHIR., F.R.C.S.

Surgeon, St. Peter's and St. Paul's Hospitals; Genito-Urinary Surgeon, The Hospital for Sick Children, Great Ormond Street and Whipps Cross Hospital

The term mega-ureter is used to describe states of chronic dilatation of the ureter for which no organic obstructive cause can be found; it implies an ignorance of the fundamental pathology and it is hoped that it will be replaced in time by a more enlightened nomenclature; at present it is easier to say what mega-ureter is not than what it is and certainly more than one disease is included under the heading. The chronic dilatations which accompany congenital malformations such as ureterocele, ureteric ectopia and congenital stricture are not, by definition, included in the category of mega-ureter, though it may well prove that similar causes of dilatation are operative in these conditions. Dilatations secondary to congenital urethral obstructions and vesical retention, on the other hand, are also excluded but show a marked difference in behaviour from the mega-ureter.

The degree of ureteric dilatation is estimated in ordinary practice from X-rays and it is important to recognize the limits of normal variation as well as different appearances which are presented by different methods of filling. The intravenous pyelogram seems to involve least disturbance of normal conditions and is probably the most satisfactory method of delineation, though it is often invalidated by the deterioration of renal function. Nevertheless, the ureter becomes wider as the urine flow increases and since the opaque medium itself produces an osmotic diuresis, the ureter will appear more dilated in the later films of the series than the earlier. In conditions of extreme polyuria, as in diabetes insipidus, the normal ureter will reach proportions comparable with the mega-ureter and yet still be capable of efficient function. In retrograde pyelography, the distending force is to some extent under the control of the operator, yet the presence of the foreign body and the stimulus of the injection result in a forceful contraction and, in most cases, a ureter will appear less dilated in the retrograde than in the intravenous series. Finally, when uretero-vesical reflux is present and ureteric filling occurs during a micturating cystogram, the distending force is very considerable; under such circumstances a ureter which is normal on intravenous pyelograms may appear grossly dilated. In assessing progress of an individual case, it is therefore vital to ensure that the films at various periods are strictly comparable.

The effects of acquired disease are not always easy to distinguish from the congenital mega-ureter; it is recognized that infection can be responsible for some ureteric dilatation, but confusion arises from the fact that mega-ureter is frequently complicated by infection. When a large calculus is present in the renal pelvis, it is common to find some dilatation of the lumbar ureter beneath it; this may be due to infection or to a disorder of ureteric activity consequent upon the pelvi-ureteric obstruction, but it is in an acquired lesion and not a form of mega-ureter. Slight dilatation of the whole length of the ureter is also a feature of many cases of congenital hypoplasia of the kidney with chronic urinary infection. Infection and cystitis with oedema of the ureteric orifice interfere with the normal valvular action of the uretero-vesical junction and reflux may therefore occur; it sometimes happens in a case of chronic urinary infection in which the intravenous pyelogram shows only minimal dilatation, that a cystogram demonstrates reflux into grossly dilated ureters. In these cases it is possible that some congenital abnormality is present, yet the prognosis appears to be good if the urine is kept sterile and no permanent dilatation is seen despite the ready distensibility during micturition.

The Mega-Ureter-Megacystis Syndrome

In one of the syndromes associated with mega-ureter, the bladder shows a characteristic disturbance of function, distinct from, though sometimes confused with, the results of urethral obstruction. The pathology of this syndrome is still unexplained and the term mega-ureter-megacystis is therefore only a convenient description. The disorder usually presents early in childhood and affects both sexes equally.

Pathology

Bilateral ureteric dilatation is present, often at birth, and may reach extreme proportions; vesico-ureteric reflux is usually free though in some it occurs only during micturition and active retrograde peristalsis has been observed in these ureters by cine-radiology. The bladder is of large capacity, but there is little evidence of detrusor hypertrophy and never more than a slight trabeculation; the bladder neck and urethra are normal. It has been claimed by Swenson *et al.* that the number of ganglion cells in the bladder wall is diminished in this condition and that it is analogous to, indeed sometimes associated with, Hirschsprung's disease. This association has not been observed in any case at the Hospital for Sick Children and in two post-mortem examples of megacystis a careful count by Dr. Leibovitz has shown a normal ganglion cell population.

Clinical, Cystoscopic and Radiological Features

The majority of children present with persistent or recurrent urinary infection, often with some impairment of renal function. In the older cases it has often been noticed that the child passes urine rather infrequently but passes a large volume at a time; moreover, on trial it will be found that a second and third micturition is possible a few minutes after the first. Radiologically it can be demonstrated that this is due to the reflux of a large volume of urine into the ureters with each bladder contraction: the bladder empties itself at each micturition, but as soon as it relaxes it is filled again from the distended ureters, leading to a 'false' residual urine. This is in sharp contrast with the true residuum found in the obstructed bladder, but unless double micturition is tried, the two are easily confused and often at first examination children with the mega-ureter-megacystis syndrome are suspected of bladder-neck obstruction. A further difficulty arises from the fact that after instrumentation or during a severe infection there may be a temporary phase of true retention.

Intravenous pyelograms show the ureteric dilatation provided renal function is adequate, cystograms show it more clearly (Fig. 1). On cystoscopy the large bladder capacity will be noted, although trabeculation is absent or slight. The ureteric orifices usually gape widely, so widely in fact that except for their obliquity they resemble the orifices of diverticula and the urethroscope can easily be introduced into them. A characteristic motility is also observed, the ureter is widely dilated at rest but exhibits from time to time a powerful contraction which, however, only obliterates the lumen at the ureteric orifice. A cystometrogram performed with slow filling will demonstrate the large bladder capacity; often a child of 6 or 7 years will hold 900-1,000 ml. and the intravesical pressure will rise only towards the end of this filling. In spite of this capacity, active contractions are still capable of emptying the bladder.

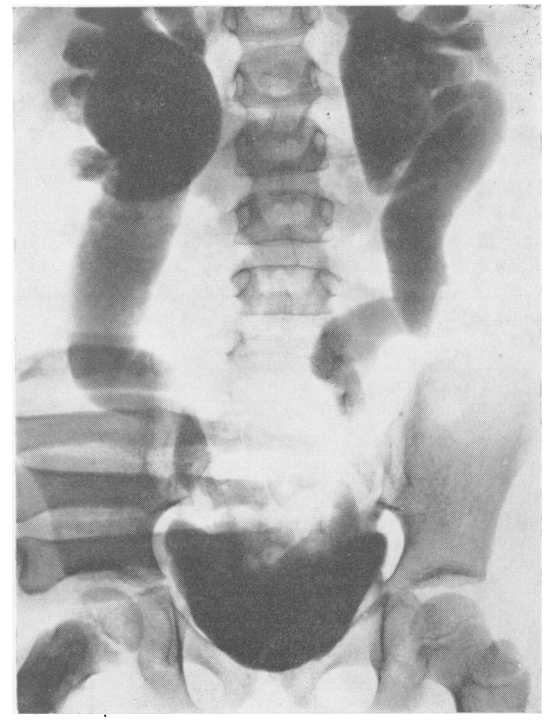

FIG. 1.—Mega-ureter-megacystis syndrome. Cystogram with reflux in a girl aged 5 years, with a long history of general ill-health, vomiting, and nocturnal enuresis. Bladder usually palpable after micturition, but cine radiology shows this to be false residual due to return of urine which had refluxed into ureters. On cystoscopy, the bladder wall was smooth, but the ureteric orifices were widely dilated. Ureteric contractions active but ineffective.

Differential Diagnosis

The differentiation from simple mega-ureter is made without difficulty from the observation of the large bladder and the gaping ureteric orifices. Congenital bladder-neck or urethral obstruction, however, can also produce an increased capacity and ultimately reflux into dilated ureters, and the following points must be noted in diagnosis: First of all, severe obstructions are common in boys but rare in girls, the megacystis affects both sexes equally and therefore a girl is more likely to be suffering from the latter. Trial of double micturition will often distinguish the true from

the false residual urine, and whereas overflow incontinence is common in obstruction it is rare in megacystis. Cystoscopy in the obstructed bladder will show heavy trabeculation and sacculation, while even if the ureteric orifices are incompetent they are usually surrounded by hypertrophied muscle and cannot become widely dilated; bladder-neck hypertrophy and some urethral lesion are also likely to be found. The bladder capacity in cystometry is not so enormous in the obstructed case as in the megacystis and pressure rises more rapidly. When an advanced dilatation of the ureter with reflux is due to obstruction, peristaltic activity is usually absent or minimal, whereas the mega-ureter retains the ability to make forceful although ineffective contractions.

Treatment

In mild cases, which are the majority, sterilization of the urine by chemotherapy and a regime of double or triple micturition suffice to prevent any further progress of the disease. The child should be induced to empty the bladder at least every three hours by day and preferably once at night, and the emptying process should always be completed by a second and, if necessary, a third micturition after each initial attempt. A single course of chemotherapy may be enough or continuous treatment with small doses may be required. Deterioration can be due to pyelonephritis, a progressive renal scarring independent of the urinary stasis or to a gradual loss of efficient bladder emptying. In the latter circumstances a bladder-neck resection or an anterior Y-V-plasty at the vesico-urethral junction may lead to an improvement and enable the double micturition regime to be carried out more efficiently. Operations designed to prevent reflux or narrow the ureters by plastic reduction are still experimental in this syndrome and not of proved value. Sympathectomy is unlikely to be helpful.

Simple Mega-Ureter

Mega-ureter associated with a bladder of normal capacity and function may be unilateral or bilateral, it occurs in both sexes and although it may be present in childhood, many are only discovered in adolescence or adult life.

Pathology

In most the whole length of the ureter is dilated down to the intramural portion which is of normal calibre. Occasionally a sudden and unaccountable narrowing occurs a short distance above the bladder, but the 'narrow' segment does not show any evident abnormality and in bilateral cases the supravesical narrowing may be present on one side only. The muscle of the dilated portion is always hypertrophied, and until the latest stages remains actively contractile. It has long been suspected that a 'functional' obstruction must be present at the lower extremity and Murnaghan has recently suggested that there is an abnormality in the distribution of muscle fibres which causes a contraction wave to be initiated at the lower end, to travel upwards and to obstruct the normal downward flow of urine.

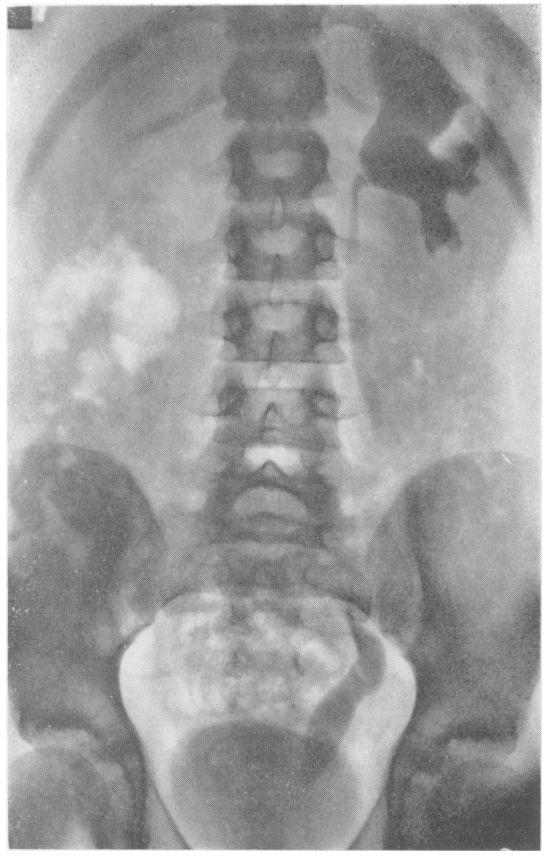

FIG. 2.—Simple mega-ureter (solitary kidney). Intravenous pyelogram on girl aged 7 years, who had suffered a single attack of urinary infection. Mild dilatation of the lower end of the left ureter. No kidney found on the right side. The condition was entirely unchanged and the child symptom-free two years later.

Clinical, Radiological and Cystoscopic Features

Most cases present with recurrent or persistent urinary infection and have only slight loin pain as a localizing symptom. In cases with sterile urine, there may be some lateral abdominal pain, but this seldom comes in such severe spasms as the pain of hydronephrosis due to pelvi-ureteric

obstruction. Haematuria has occurred as an isolated symptom in some, and although this may suggest the presence of a stone it is not always possible to find one. Nevertheless, secondary calculi in the dilated segment are relatively common in both sterile and infected cases.

The intravenous or retrograde pyelograms demonstrate the ureteric dilatation for which no other causes can be found. The bladder is normal on cystoscopy and cystometry, and the ureteric orifice is either entirely normal or appears to be raised on a small pyramidal eminence. Although this appearance may be confused with that of a ureterocele, it is quite distinct and more in the nature of a slight prolapse which can in fact be reduced at operation by pulling on the ureter. A ureteric catheter passes easily into the orifice and carefully directed X-ray films will show whether the dilatation reaches down to the bladder wall or ceases above (Fig. 2). Vesico-ureteric reflux is not usual in these cases and it is never as free as in the mega-ureter-megacystis syndrome, but a little regurgitation may occur during micturition.

Differential Diagnosis

Congenital or acquired stricture is distinguished by the obstruction to the passage of a ureteric catheter and most other congenital ureteric anomalies produce distinctive radiological and cystoscopic appearances. In bilateral simple mega-ureter, the normality of the ureteric orifices and normal bladder capacity differentiate from mega-ureter-megacystis. When stones are present, it is important to judge whether they are primary or secondary; in the mega-ureter a complicating calculus is usually rounded and freely mobile up and down the ureter.

Treatment

In most cases, whether unilateral or bilateral, the urine can be sterilized by chemotherapy and often no further treatment is required. Such cases have been carefully followed up for ten years and over, and have neither suffered any symptoms nor shown any increase of dilatation.

In advanced unilateral cases with renal damage and possibly calculi as well, nephro-ureterectomy is the treatment of choice.

In bilateral cases where there is persistent infection, increasing dilatation or stone formation, and in a few unilateral cases with similar signs but a well-preserved kidney, the uretero-vesical junction should be excised and the ureter re-implanted into the bladder in such a manner as to prevent uretero-vesical reflux. This is best accomplished by turning the severed end back on itself for 1-2 cm. and implanting it high into the bladder with this projecting nipple (Fig. 3). Where there is a supravesical narrow segment, the same technique may be employed or the actual cone and site of the narrowing may be excised and the two ends of the ureter rejoined by an end-to-side anastomosis.

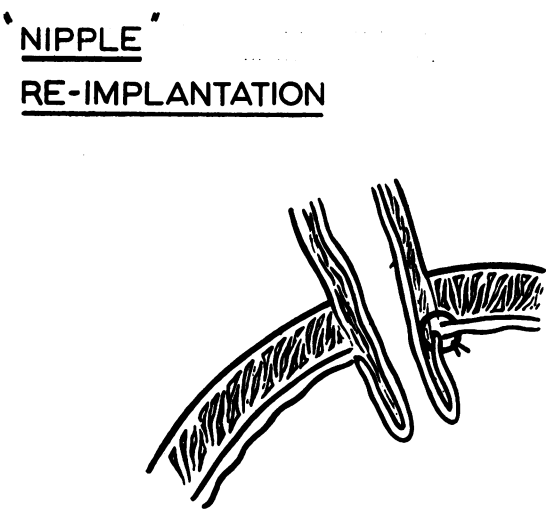

FIG. 3.—Diagram to show the method of 'nipple' re-implantation of the ureter into the bladder.

Plastic reduction and straightening of the kinks in a very dilated and tortuous ureter may be performed at the same time as the re-implantation or an enormous organ may be better replaced by a loop of ileum.

When there is any suggestion of a stricture at the lower end of the ureter, instrumental dilatation is worth a trial.

BIBLIOGRAPHY

WILLIAMS, D. I., BODIAN, M., EDWARDS, D., LEIBOVITZ, S., and MURNAGHAN, G. F. (1957), *Brit. J. Urol.* **29**, 389.

SWENSON, O. (1955), *Arch. Dis. Childh.*, **30**, 1.