Neuropathic Bladder

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Almost all patients with neuropathic bladder suffer urinary incontinence and an appreciable proportion also stand at risk of secondary upper renal tract complications, potentially leading to end-stage renal failure [1]. Management encompasses both problems and aims to reproduce, so far as possible, a normally functioning bladder, namely one with adequate low-pressure functional capacity, yet which can be emptied completely when wished. Spinal neurological disease can adversely affect storage or voiding functions (and quite often both together) in several ways and a description of the pathophysiological processes involved is necessary before considering treatment.

PATHOPHYSIOLOGY

General

Knowledge of neuropathic bladder dysfunction owes much to urodynamics, particularly video-urodynamics, techniques which were first applied to adults with cord trauma [2] and subsequently to children with congenital lesions [3]. Contrary to common belief, the patterns of dysfunction demonstrated urodynamically are common to both groups of patients though do differ in proportion between the two.

Classification

Spinal innervation of the bladder and its sphincters exists principally within the 2nd to 4th sacral segments (conus medullaris) and classification of neuropathic bladder has traditionally depended upon the site of the cord lesion in relation to the conus.

With suprasacral lesions (hyperreflexic bladder) the conus is intact, but isolated from higher centres; the bladder and sphincters retain their spinal innervation so that voiding occurs only by sacrally mediated reflex detrusor contractions. This form is recognisable clinically by positive conus reflexes (ano-cutaneous, glans-bulbar).

Sacral cord lesions destroy the innervation of the detrusor and sphincters (areflexic or autonomous bladder); there is no detrusor contractibility and voiding occurs by overflow or by raising intra-abdominal pressure. Conus reflexes are negative.

The distinction between these two patterns is determined by the caudal extent of the lesion; the cranial extent and hence the primary neurological level, is immaterial.

It is now appreciated that traditional classification does not encompass all aspects of neuropathic bladder dysfunction and that when assessing a patient urodynamically "classification" is not the principal concern which is rather to determine whether the problem is one of storage or voiding malfunction and the reason or reasons for this. Nonetheless, for present purposes, the traditional classification is retained.

Hyperreflexic bladder (Fig. 1)

Essential to this form of dysfunction is that the sphincteric mechanism, usually including the bladder neck [4], is competent in the face of any rise in intra-abdominal pressure. At a certain point, usually fairly constant in any individual, filling induces a reflex detrusor contraction (hyperreflexia). The same sensory input also triggers a mass pelvic reflex so that there is almost always detrusor-sphincter dyssynergia. Initially this is complete but, after a variable time, the external urethral sphincter par-

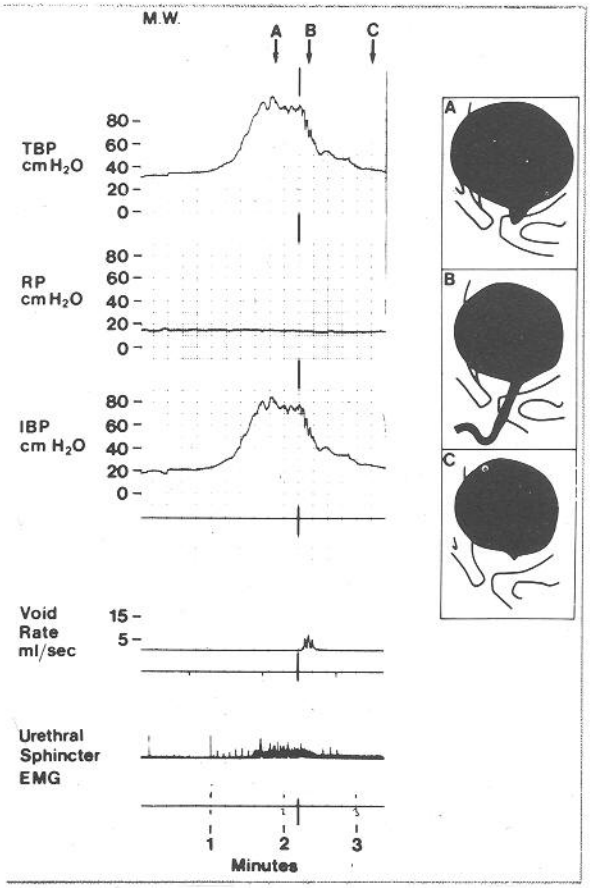


Figure 1. Hyperreflexic bladder (top trace total bladder pressure, second trace rectal pressure, third trace intrinsic detrusor pressure). Voiding occurs solely by a reflex detrusor contraction with initally complete detrusor-sphincter dyssynergia (A). Voiding occurs as the external urethral sphincter partially relaxes (B). Although generating high pressure, the detrusor contraction fades away before the bladder is empty (C) (non-sustained contraction).

tially or wholly relaxes and voiding proceeds. As a rule this partly obstructed voiding leads to high-pressure detrusor contractions but these tend to fade away before the bladder is empty (non-sustained contractions).

Areflexic (Autonomous) bladder (Fig. 2,3)

There are no detrusor contractions and voiding occurs by overflow or by increasing intra-abdominal pressure. Thus there is always some degree of sphincter weakness incontinence (SWI) though this varies widely and unpredictably from one patient to another. In boys any element of outflow obstruction is located at the level of the external urethral sphincter (Fig. 2) whereas in girls the entire sphincteric zone tends to be involved. Unlike detrusor-sphincter

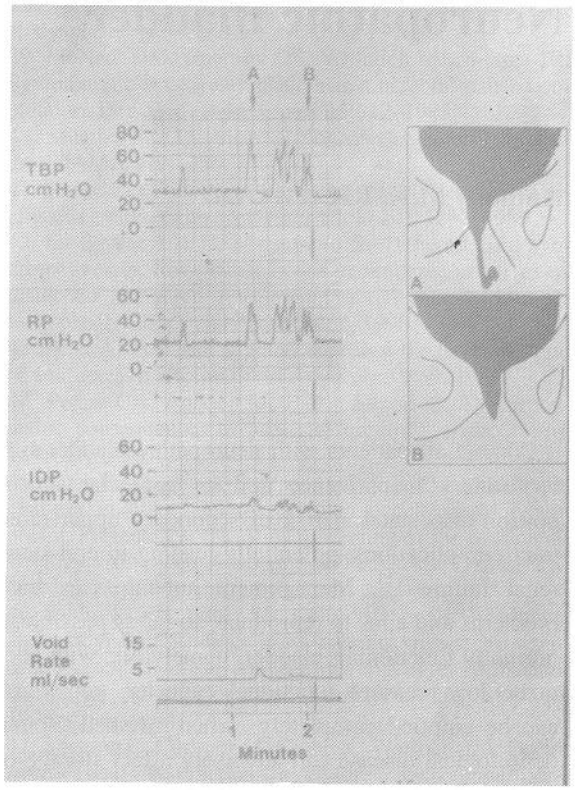


Figure 2. Areflexic bladder. Voiding occurs by raising intra-abdominal pressure (A). During voiding there is partial obstruction at the level of the external urethral sphincter (static sphincteric obstruction) which ultimately becomes complete (B).

dyssynergia, which is a dynamic phenomenon, such obstruction constitutes a **fixed** urethral resistance (static sphincteric obstruction) [4]. Those with high resistance have good functional bladder capacity at the expense of large residual urine (Fig. 3); those with gross SWI have no residual urine but lack any useful functional bladder capacity (Fig. 3).

Intermediate bladder (Fig. 4,5)

In this form, outwith the traditional classification, reflex detrusor contractility, usually generating only modest pressures, is combined with some degree of SWI so that voiding occurs by two means (Fig. 4). As a rule, any outflow obstruction resembles static sphincteric obstruction rather than detrusor-sphincter dyssynergia. Conus reflexes are negative.

Among myelomeningocele patients, some 65 % have intermediate bladder, 25 % hyperreflexic bladder and 10 % areflexic bladder. With other congenital cord lesions, the proportion of patients with hyperreflexic bladder is appreciably smaller.

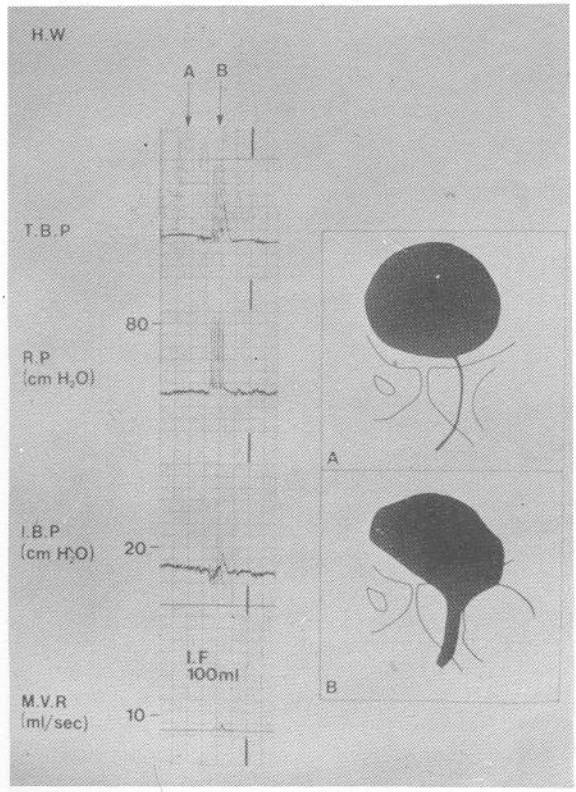


Figure 3. Areflexic bladder (contrast-filled urethral catheter in situ). At rest the bladder neck is closed (A) but the entire sphincteric mechanism becomes grossly incompetent when intra-abdominal pressure is raised by coughing (B).

Detrusor non-compliance (Fig. 5)

Normally baseline intravesical pressure rises only imperceptibly within the physiological range of capacity. In patients wih neuropathic bladder, a rise in baseline pressure due to detrusor non-compliance is common, especially with intermediate dysfunction, and this may proceed steadily (Fig. 5), or may occur early or late in the filling phase.

Where there is appreciable SWI, non-compliance serves to further compromise functional bladder capacity (Fig. 5). In the presence of marginal SWI, or a fully competent sphincteric mechanism, non compliance lead to high intravesical pressure and, in turn, to secondary ureterovesical obstruction or reflux [5]. These complications are apt to occur whenever baseline intravesical pressures persistently exceed 20 cm H_2O.

Non-compliance is of unknown cause but is usually accompanied by an increase in the usual collagen/muscle ratio within the detrusor. Thickening

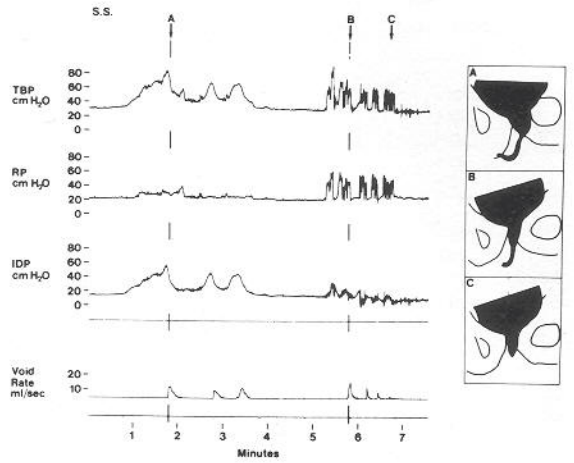


Figure 4. Intermediate bladder. Voiding occurs both by detrusor contractions (A) and, because there is sphincteric incompetence, by raising intra-abdominal pressure (B). Residual urine exists because of static sphincteric obstruction (C).

and sacculation of the bladder wall (Fig. 6) are typically associated with non-compliance rather than with high-pressure detrusor hyperreflexia.

Incomplete cord lesions

A few patients with neuropathic bladder achieve spontaneous, if precarious, urinary continence. All have incomplete cord lesions, with sacral sensory sparing (occasionally motor sparing also) and most have a combination of positive conus reflexes and spinothalamic (pain, temperature) sacral sparing. Marked urgency of micturition is characteristic of these patients and, despite their continence, they remain at risk of upper renal tract complications.

Table 1. Factors in storage and voiding failure in the neuropathic bladder

Storage failure
Detrusor hyperreflexia
Detrusor non-compliance
Sphincter Weakness Incontinence (SWI)
Voiding failure
Detrusor-sphincter dyssynergia
Static sphincteric obstruction
Non-sustained detrusor contractions

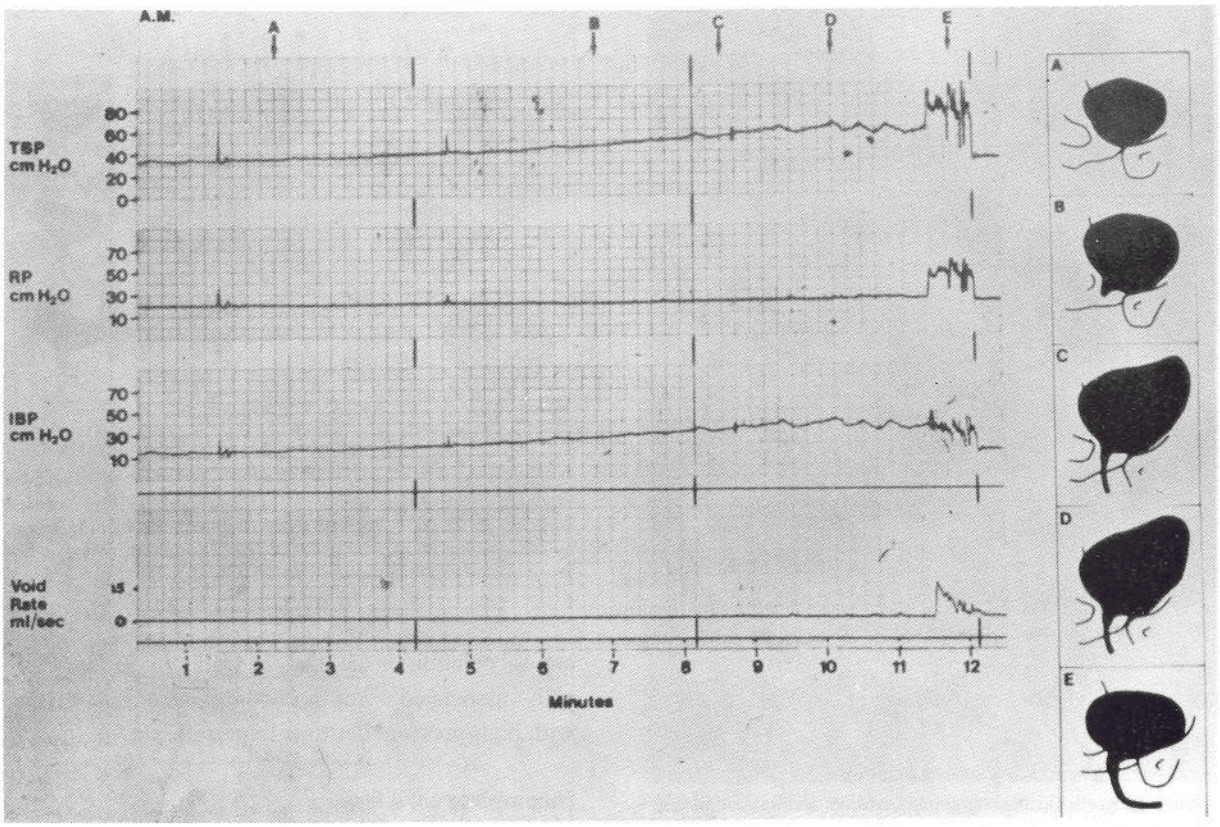


Figure 5. Intermediate bladder. Initially the bladder neck is closed (A) but passive opening occurs as intravesical pressure rises during bladder filling (B). With a further rise in baseline pressure (detrusor non-compliance) there is overflow incontinence (C) with further leakages occurring as a result of low-pressure detrusor contractions (D). The bladder is emptied by abdominal straining (E) and baseline intra-vesical pressure returns to normal.

Urinary storage and voiding

Factors compromising storage and voiding in the neuropathic bladder are summarised in the Table 1. In patients with hyperreflexic bladders any lack of capacity is almost always solely due to reflex detrusor contractions, whereas in those with areflexic or intermediate dysfunction capacity may be impaired by one or more factors.

Upper renal tract complications

These are more common in children with all types of cord lesion than in adults with cord trauma and, unlike the latter, occur in females too, albeit less often than in males. Periods of greatest risk are infancy and puberty though no age is exempt, including on into adult life.

Upper renal tract complications are rare in the absence of bladder outflow obstruction and never occur where the bladder is permanently empty due to gross SWI.

In patients with neuropathic bladder, outflow obstruction is usually associated wih appreciable residual urine. Nonetheless, normal upper renal tracts are often maintained in the presence of a large residual urine and it is now appreciated that upper tract complications occur only where outlet obstruction is compounded by high intravesical pressures caused by detrusor non-compliance or hyperreflexia [6].

The importance of urinary infections as a cause of renal damage in patients with neuropathic bladder has been exaggerated. They are clearly of potential significance in the presence of vesicoureteric reflux, but their occurrence otherwise is indicative of disordered hydrodynamics within the urinary tracts. Once these have been corrected the infections resolve, often leaving no residual renal damage.

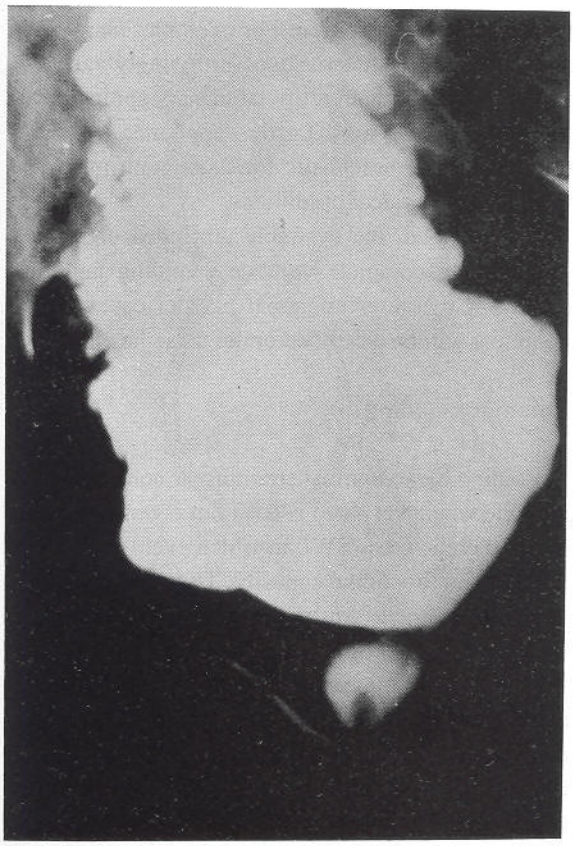


Figure 6. Cystogram of a patient with a non-compliant neuropathic bladder. The detrusor shows typical sacculation. The bladder neck is incompetent but there is outflow obstruction due to static sphincteric obstructon.

PATIENT ASSESSMENT

Continuous dribbling of urine, or leakage during coughing, crying, etc., is indicative of SWI as also is "expressibility" of the bladder. Patients with hyperreflexic bladders tend to void spontaneously at discrete intervals, often with an interrupted stream.

Neurologically, presence or absence of conus reflexes should be determined and so also any sacral sensory or motor sparing. No matter how slight the neurological deficit otherwise, neuropathic bladder is certain in the absence of conus reflexes or of sacral sensory sparing [7]; both signs are demonstrable neonatally.

Imaging studies

The state of the upper renal tracts is most conveniently monitored by serial ultrasound examinations supplemented, as necessary, by radionuclide studies. Ultrasound also finds use in estimation of residual urine and bladder wall thickness; an increase in either often presages upper renal tract complications.

Urodynamic examination

This is best combined with simultaneous videoradiological examination [2] and is ideally performed by the clinician also responsible for the patients' care. Per-urethral catheters are routinely used and slow filling is employed to reproduce, so far as possible, the bladder's normal behavior. (The common patterns of dysfunction are illustrated in Fig. 1-5).

Urodynamic examination may be indicated when upper renal tract complications occur or when planning treatment for incontinence. The study may also be conducted neonatally to assess future risk of upper tract complications [8]. However, bladder function may change in time, almost always for the worse [9], so that a "safe" bladder neonatally, or at any other time, is no guarantee of freedom from future risk.

Assessment without urodynamics

Where urodynamic facilities are lacking, useful information concerning bladder function can be gleaned by other means. Thickening of the bladder wall, as detected by ultrasound, is indicative of detrusor non-compliance. Bladder dysfunction is predictable in the presence of positive conus reflexes when any lack of functional capacity is due to detrusor hyperreflexia. In the absence of conus reflexes, residual urine should be measured by catheterisation. A large residual indicates that there is minimal SWI and continence is likely with intermittent self-catheterisation alone. Small residual urine volume is indicative of gross SWI requiring surgical correction to achieve continence.

MANAGEMENT

General

The basic principles are that:

- maintenance of renal function takes precedence over treating incontinence
- treatment should relate to the nature of the bladder dysfunction

• because dysfunction is often complex, individual forms of treatment must often be used in combination
• management should **realistically** relate to the patients physical and intellectual status generally.

The last deserves elaboration. Patients with neuropathic bladder are rarely untroubled by other disabilities. Neuropathic bowel is almost universal and often merits treatment ahead of the urinary incontinence. Some degree of paralysis is usual and influence definition of "adequate" functional bladder capacity; for a patient with good mobility a capacity ensuring dryness for 2 hours is adequate, if not ideal, whereas for one wheelchair-bound at least twice that capacity is desirable. Complete paraplegia is frequently compounded by major spinal deformities which may make self urethral catheterisation impossible for girls or render siting of stomas, including continent stomas, problematical for both sexes. Lastly, and perhaps most importantly, myelomeningocele, still much the commonest cause of neuropathic bladder in children, is accompanied by hydrocephalus and the effects of this malformation upon intellect and manipulative skills may make advanced forms of treatment unrealistic.

Management during infancy

Here attention concentrates upon the upper renal tracts, preferably in a manner not compromising future continence. "Prophylactic" intermittent catheterisation is advisable, where practicable, in infants with "unsafe" bladders [10] and antibiotic prophylaxis in those with vesicoureteric reflux.

Established complications of moderate degree may respond to a regime of intermittent catheterisation. More severe problems are best handled by a temporising cutaneous vesicostomy [11] which is maintained until the child is old enough for more definitive treatment.

Management of older children

General

Here appliance-free urinary continence becomes an additional goal and the age at which this is sought varies according to the child's abilities generally and to the treatment proposed. For example, many children learn self-catheterisation surprisingly early but few cope well with an artificial urinary sphincter before 8 years of age. Lastly, appliance-free continence may be unrealistic for those with major intellectual or physical disabilities.

As described, the desirable attributes of bladder function are complete voluntary voiding and adequate low-pressure functional bladder capacity and treatment will be described under these headings.

Voluntary voiding

Voiding by abdominal straining or compression is possible whenever there is SWI but is only complete where there is gross SWI, in which event the bladder lacks useful functional capacity. Thus this method of voiding is largely restricted to patients managed with an artificial urinary sphincter.

Clean intermittent self-catheterisation (CISC) is applicable to all forms of dysfunction but will secure continence only if the bladder has adequate functional capacity and will maintain the upper renal tracts only if that capacity is reached without excessive intravesical pressure. Thus CISC must often be combined with measures to secure adequate, low-pressure, functional capacity.

Capacity impaired by detrusor hyperreflexia

Detrusor hyperreflexia usually responds to the antispasmodic oxybutynin. Side-effects are unusual in children but, where troublesome, can be overcome by instillation of the drug intravesically, in standard dosage, during episodes of CISC. Augmentation cystoplasty is necessary where hyperreflexia does not adequately respond to oxybutynin.

Capacity impaired by detrusor non-compliance

Although occasionally responsive to drugs directly inhibiting the detrusor (imipramine, oxybutynin), non-compliance usually demands augmentation cystoplasty. This is normally effected by the ileal clam patch technique (Fig. 7) although a pouched cystoplasty, using small or large bowel, is advisable where the bladder is small (Fig. 8). Creation of a low-pressure reservoir by cystoplasty may overcome minor degrees of SWI whilst with

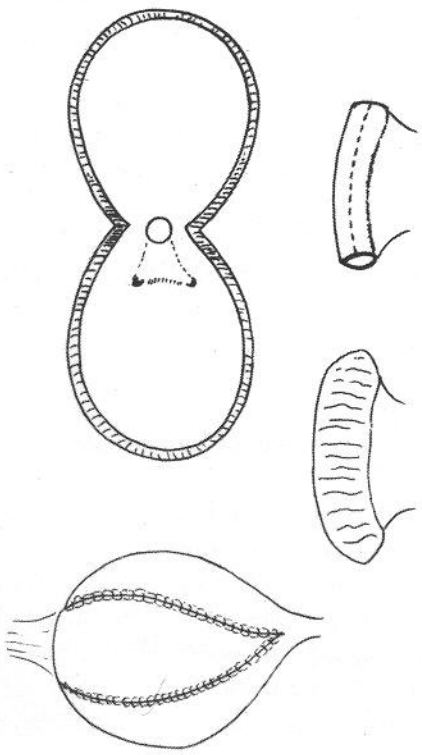


Figure 7. Ileal Clam-Patch Cystoplasty. The bladder is opened, in the coronal plane, from dome almost to the bladder neck. An ileal segment, of appropriate length, is detubularised and then anastomosed, as a patch, to the opened bladder.

somewhat more pronounced SWI it may be necessary to insert only the cuff of an artificial sphincter rather than the entire device [12].

Capacity impaired by SWI

Minor degrees sometimes respond to alpha-adrenergic agonists (e.g. ephedrine). Major degrees require surgical correction and no method currently available guarantees success.

For boys the artificial urinary sphincter is usually employed (Fig. 9), preferably with cuff placement around the bladder neck rather than the bulbar urethra (the latter is only practicable post-pubertally). The device is expensive and patient selection must be rigorous. Essential prerequisites are adequate intelligence, motivation, manipulative skills and mobility, and upper renal tracts which are ideally normal or at least stable. Any detrusor hyperreflexia or non-compliance not controllable by medication de-

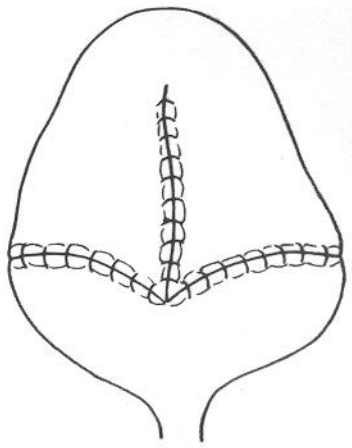


Figure 8. Pouched cystoplasty. The bladder is opened in the sagittal plane. A detubularised segment of bowel is formed with an inverted "U" pouch and anastomised to the opened bladder.

mands simultaneous augmentation cystoplasty [12]. With gross SWI voiding is entirely by abdominal straining; with lesser SWI this can be supplemented by CISC or, alternatively, endoscopic sphincterotomy can be performed to enable complete voiding by straining. Where the entire sphincter device has been inserted there is an appreciable need for revisionary surgery because of component failure or disconnection and, in non-augmentated cases, development of detrusor hyperreflexia or non-compliance [13] may lead to upper renal tract complications.

Although the artificial urinary sphincter can be used for girls, current preference is for procedures deliberately aiming to create bladder outflow obstruction and with voiding entirely by CISC. Techniques employed include bladder neck repair or suspension [14] (Fig. 10), sling procedures and the Kropp neourethroplasty [15]. Published results do not suggest that any of these methods is inherently superior and all should be combined with augmentation cystoplasty if there is any element of detrusor hyperreflexia or non-compliance [14].

Management of vesicoureteric reflux

Treatment of this common problem by operative reimplantation in the presence of untreated bladder outflow obstruction or detrusor hyperreflexia or non-compliance invites complications in the form of secondary ureterovesical obstruction or development of contralateral reflux [16]. By contrast, with

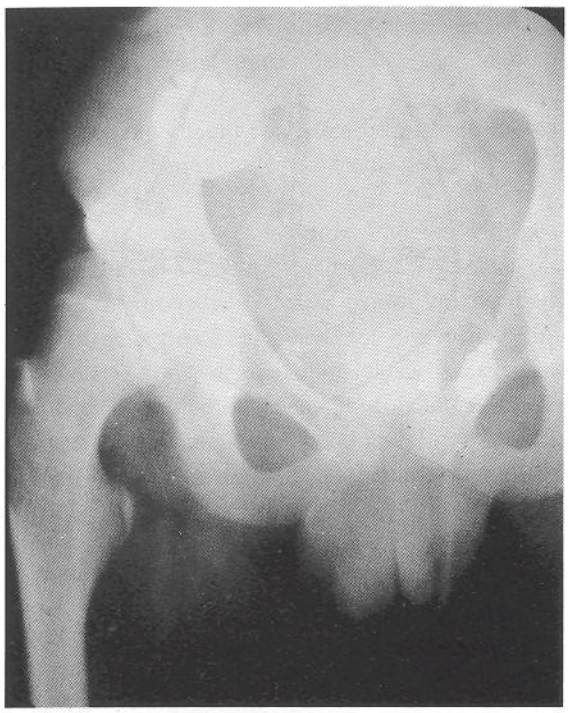
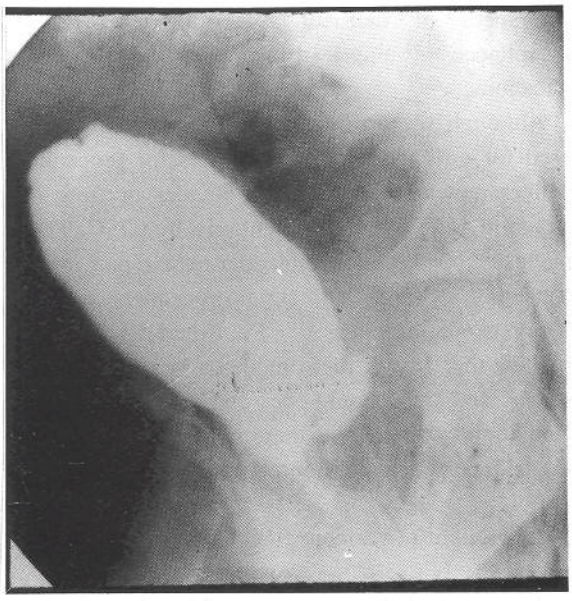


Figure 9. AMS 792 artificial sphincter in situ. The cuff is placed around the bladder neck. The pressure-regulating balloon lies in the right iliac fossa. The control assembly is situated in the left groin and the pumping chamber, which empties the cuff, lies at the base of the left hemiscrotum. In the current, AMS 800, device the pumping chamber and control assembly are combined as an integral unit.



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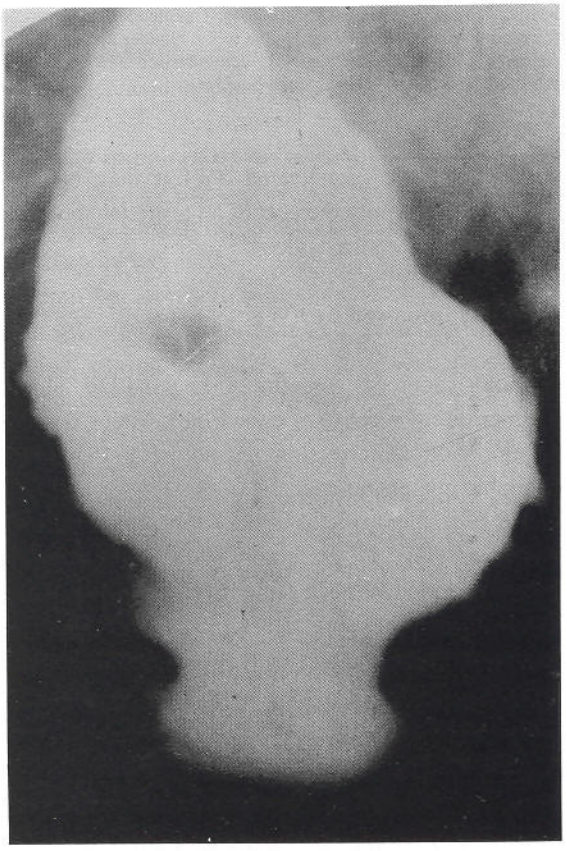


b

Figure 10. Female patient with neuropathic bladder. The preoperative cystogram (a) shows a grossly incompetent sphincteric mechanism (with reflux into the stump of a urethrally ectopic ureter). The detrusor was non-compliant. Cystogram following combined bladder neck suspension and pouch augmentation cystoplasy (b). The bladder neck is competent and the pouch cystoplasty is seen superiorly and the native bladder inferiorly.

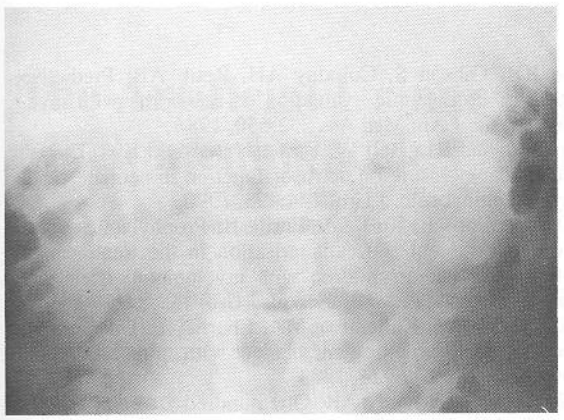
treatment of the bladder disorder itself, reflux usually resolves [17] or becomes inconsequential.

Results

With modern management upwards of 80 % of children can be rendered reliably dry by day with normal or stable upper renal tracts [18,19]. In approximately half this is achievable by CISC alone or with adjunctive medication. The remainder require surgery and most unsatisfactory results are due to failure to correct SWI.

Management of the severity disabled

Here the goal of appliance-free urinary continence is often unrealistic. Most boys can be managed with a penile appliance, plus endoscopic sphincterotomy if there is any bladder outflow obstruction (Fig. 11). Indwelling urethral catheterisation, if properly handled, represents a useful option for girls [20]. Where these methods fail, or are unacceptable, there re-



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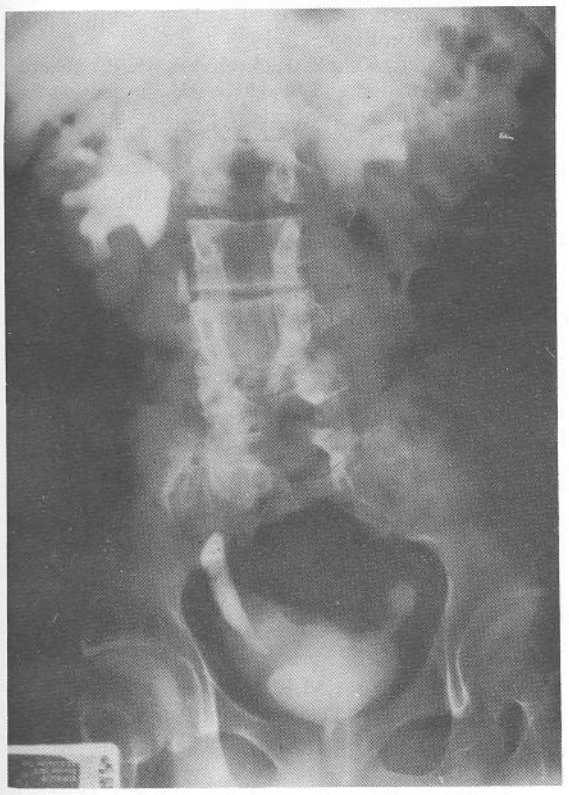

b

Figure 11. Male with neuropathic bladder. An IVU (**a**) shows faint nephrograms bilaterally in grossly hydronephrotic poorly functioning kidneys. Three months post-endoscopic external sphincterotomy a further IVU showns substantial resolution of the upper renal tract changes, (**b**). Ideally sphincterotomy should be undertaken before such complications occur.

mains a role for non-continent urinary diversion, preferably by way of a non-refluxing sigmoid conduit.

For children with major physical disabilities yet well preserved intelligence and manipulative skills, continent diversion is more appropriate, usually employing the Mitrofanoff principle [21]. The same approach is sometimes necessary for those with

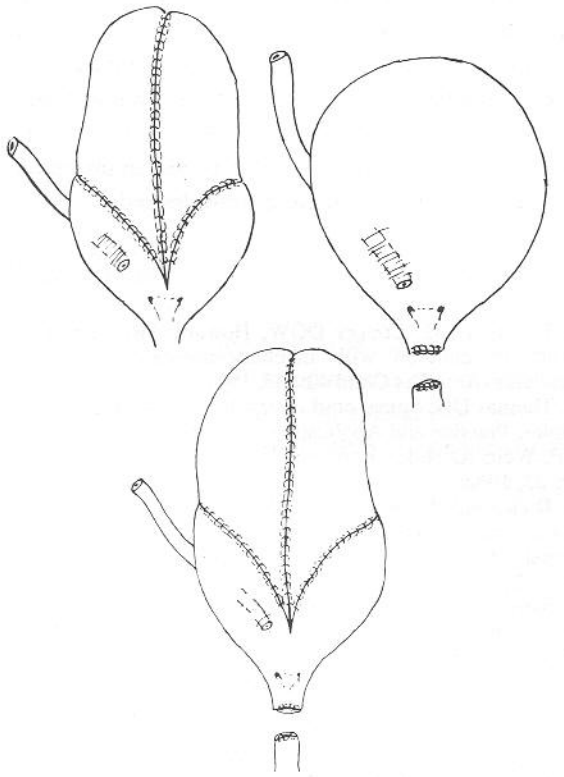

Figure 12. As described in the text, in some circumstances the Mitrofanoff procedure may need to be combined with augmentation cystoplasty (top left), with occlusion of the bladder neck (top right) or, occasionally, with both (bottom).

minor physical disabilities unable to practice CISC because of intact urethral sensation associated with an incomplete cord lesion. As a rule this problem is confined to boys, particularly those with sacral agenesis or neuropathic bladder associated with imperforate anus. As with other means of treatment, the Mitrofanoff principle demands adequate low-pressure functional bladder capacity obtained, if necessary, by appropriate medication or by augmentation cystoplasty. In the presence of SWI the bladder neck may require surgical occlusion (Fig. 12) with the disadvantage that this leaves limited access to the bladder in the event of complications such as stone formation.

CONCLUSION

Despite advances in recent years, it should not be forgotten that neuropathic bladder is a treatable, not curable, conditon and that the history of its treatment

has been and remains one of successive expedients. Current expedients are both more scientifically based and more effective than those of times past, but expedients they remain with all that implies. With the arguable exception of CISC, urinary continence demands a price either in the form of long-continued medication or comparatively major surgery, both with known side-effects and an uncertain future [21]. There is still much to be desired.

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