Bladder pressure and kidney function in children with myelomeningocele: review article

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Therapy of children with myelomeningocele should achieve a double goal: on the one hand, obtain continence and, on the other hand, preserve kidney function without complications. The first goal is a social and psychological one, the latter is a vital one.

Key words: myelomeningocele; urinary bladder hyperpressure; neuropathic incontinence.

Bladder function

Capacity and residual urine

In order to obtain continence a sufficient bladder capacity is necessary. In the literature the bladder capacity values in normal children vary widely according to different authors. The values of Muellner,¹ Starfield² and Berger *et al*³ are rather similar, while the capacities mentioned by Van Gool *et al*⁴ and Esperanca⁵ are much lower and those of Hallman⁶ and much higher than the values of previous authors (Fig 1). The reasons underlying these discrepancies are numerous. The maximum capacity determined during the urodynamic session is different when the patient is asked to suppress the micturition act as long as possible and when he is allowed to void at the time of the first uncomfortable sensation. The psychological stress of a child who wishes to finish the examination as soon as possible is different from that of a quiet, eventually mentally retarded child, or a child drinking milk or under superficial anaesthesia. After water load the capacity is smaller than with physiological filling and the bladder filling rate can therefore also play a role; girls seem to have larger capacities than boys.⁵

For a long time a residual urine of less than one-sixth of the bladder capacity has been considered to be the golden standard in urological bladder disorders. However, this theoretical value must in practice be adapted to the particular situation. For

example, if there is infected urine, these residues can be detrimental to the bladder and the kidney, while larger residual volumes are acceptable if sterile.

Post voiding residual urine expressed as a percentage of bladder capacity is related to the cystometric pattern: in hypertonic dys-synergic bladders the post voiding residual urine is 65% against 9% in synergic bladders, while in hypotonic bladders the post-voiding residue is about 25% in dyssynergic hypertonic as well as in synergic bladders.⁷ High residual urine is important in different

Figure 1 Bladder capacities in children (different authors).

aspects: (1) it diminishes the functional capacity (total capacity minus residual volume), thereby increasing the frequency of micturition and the incontinence; (2) residual urine is often a result of incomplete bladder contraction at higher filling volumes. As soon as the small voided volume is replaced by diuresis, a new contraction (with leakage) occurs. In a 24 hour ambulatory measurement in a 12 year old spina bifida child, we calculated a total duration of 7.45 hours, during which the bladder pressure exceeded 40 cm of water. This total time was reduced to 1.05 hours simply by 5 intermittent catheterisations of a mean of 120 ml residual urine; and (3) regular catheterisation can improve continence as well as urinary infection, and is more acceptable to parents and children (who usually have no pain sensation). Simultaneous administration of oxybutinin gives an increased bladder capacity and low pressure.

Incontinence

Incontinence is always the result of an imbalance between intravesical pressure and urethral resistance. This relation can only be evaluated by urodynamic examinations. It should be stressed that the result of a urodynamic investigation may be different from that which would be expected from the anatomical spine defect level,⁸ but also from the clinical neurological finding such as a bulbocavernosus reflex, anal reflex, level of skin sensation and the control of the striated muscles in the lower limbs. Only urodynamic examination can detect candidates at risk of deterioration of bladder function. Although no evolution is usually expected

before the age of 2 or 3 years, this detection is already necessary in neonates especially in the presence of vesicoureteral reflux.

Bladder hyperactivity

The hyperactive bladder can be either hyperreflexive or of low compliance (Fig 2). In the first case, the baseline remains at low pressure but superimposed uninhibited contractions at variable filling volumes and pressure amplitude occur. This hyperreflexive bladder type is rather rare in its pure form in myelomeningocele patients. In those with low compliance, the pressure volume curves show a slope in which the angle is a function of the distensibility of the bladder wall. Low compliance can be due to muscle hypertrophy, denervation, fibrosis or chronic infection. According to Bauer⁹ *et al*, of the initial 50% of myelodysplastic newborns with an upper motor neuron type of neuropathic bladder dysfunction, 30% remain after puberty suggesting a progressive lesion in some children.

Even the hypoactive bladder, which in fact is areflexive and can lead in some cases to complete retention, may mask a latent hyperreflexia. A bladder which is empty most of the time because of low urethral resistance, can show a hyperreflexive pattern as soon as the outlet resistance is increased, eg after implantation of an AMS 800 sphincter or by bladder neck suspension. Detrusor hypertonia can be misjudged if the urethral resistance is so low that leakage of urine is already occurring in the presence of low bladder pressures, or when unchanging urethral resistances become equilibrated by an autonomous increasing intravesical pressure.

Figure 2 Dyssynergia between anal sphincter (AS EMG) and detrusor pressure (upper curve) during an uninhibited bladder contraction. Note also the previous abnormal decrease of activity (accompanied with mid urethral pressure fall).

Woodside^{10,11} described a simple urodynamic test which permits prediction of the final outcome of urethral resistance augmentation either by surgery or by artificial sphincters. He repeats the cystometry with a balloon catheter occluding the bladder neck so that no urine can escape; often low compliance bladders are then converted to high pressure bladders.

Urethral hyperactivity

In order to assure continence the urethral resistance must be more than 60 cm H₂O even in areflexive bladders: in this case intermittent catheterisation is the ideal solution and the patients are dry between successive catheterisations. In myelodysplasia a closure pressure of more than 60 cm H₂O occurs in only 30% of patients; this closure pressure usually decreases with time.

A hyperactive urethra can be caused by involvement of either smooth muscle or of the striated perineal musculature. Normally the urethra is closed except during voiding. Changing position from supine to sitting can cause a sympathetic dyssynergia even with inactive somatic EMG curves. According to Mundy *et al*¹² the bladder neck either competent or incompetent is never obstructive and is always incompetent in acontractile bladders at their usual bladder volumes.

Upper motor neuron lesion dyssynergia is a dynamic phenomenon which increases with age. Striated urethral muscle spasms can result with time in retractile fibrosis. Dyssynergia can be seen on permictional urethrography as a constricted area (Fig 4). During rehabilitation voiding manoeuvres (Credé or Valsalva) increased activity in the perineum can be observed; this activity counteracts the effect of these procedures.

Different types of external sphincter dyssynergia have been described:¹³ a full contraction of the sphincter throughout the increase in bladder pressure (Fig 2), a small contraction only at the beginning of the increase in bladder pressure, a waxing and waning pattern corresponding to changes in the intravesical pressure, and a sphincter hyperactivity with detrusor areflexia as seen in patients after muscarin blockade.¹⁴ It is difficult to quantify the effect of electromyo-

graphic activity on outflow resistance. By applying the technique of electromyographic differentiation¹⁵ we found in most patients a correlation of more than 0.8 between urethral pressure and electromyographic activity (Fig 3). Therefore dyssynergia may indeed be a cause for difficult voiding and thus a cause of bladder hyperpressure.

Outflow obstruction in the form of detrusor sphincter dyssynergia can also lead to impaired urinary tract drainage in a significant number of babies. It also allows identification in the newborn who are at risk from such a complication. In older children voiding dysfunction from external sphincter hyperactivity can be reflected in the simple radiographic sign of prostatic reflux, although this reflux can also occur without any sphincter activity (Fig 4). The non relaxing type of urethral hyperactivity often occurs in lower motor neuron lesions where dysuria is expressed by a static continuous urethral pressure together with an areflexive bladder.

In the literature there is disagreement about the percentage of different types of bladder-urethra behaviour. Hyperactive sphincters usually occur as frequently as areflexive sphincters. A hyperactive sphincter is associated with a hyperactive bladder in 34% of patients, whilst an areflexive sphincter occurs together with an areflexive bladder in 38% of patients. These figures by Van Gool *et al*⁴ are confirmed by Webster *et al*¹⁶, Light *et al*¹⁷ and Wijn-dale.¹⁸ The majority of patients without hyperactivity have a lesion located higher up in the spinal cord according to Naglo,¹⁹ but we could not confirm this finding.⁸

Since urethral hyperactivity provokes outflow obstruction, internal urethrotomy can be helpful.²⁰ The distant sphincter mechanism is nearly always dynamically obstructive in children with contractile bladders whereas in children with acontractile bladders some degree of static distal sphincter contraction and sphincter weakness incontinence exists simultaneously.

Evolution

Bladder function may change with time. We have already mentioned a conversion of

Figure 3 Electromyographic activity in the periurethral striated muscle (EMG in μV), urethral pressure (pressure 1000 = 50 cmH₂O) and Differential of EMG (DEMG) during a 6 second period. Note the degree of correlation (0.87).

areflexia to hyperreflexia after the implantation of an artificial sphincter, probably because the bladder wall distension increases afferent input thereby stimulating detrusor activity. The opposite situation may also occur: eg in patients undergoing sphincterotomy a hyperreflexive bladder can convert into a nearly areflexive state.

Bladder function deterioration can be ascribed to 3 different factors: (1) evolution of the central neural lesion: resorption of haematoma, fibrosis, surgical interventions, tethered cord and other evolution lesions; (2) changes in the peripheral neural system. Sundin *et al*²¹ demonstrated that in the denervated bladder a gradual increase of the orthopathic system influence occurred because of the spread of the orthopathic

nervous fibres into muscles previously innervated by the parasympathetic system. Since the orthosympathetic system augments the tone of the bladder, increased bladder pressures and low compliance occur. The beneficial effect of alpha-adrenergic blocking agents confirms this theory; and (3) fibrosis and infection provoke an increase of collagenous tissue between the muscle bundles thus decreasing the compliance of the detrusor and promoting incontinence.

Kidney function

The main reason for the follow up of patients with myelomelic is the preservation of kidney function. Indeed, vesicoureteral reflux, infection, and hydronephrosis

Figure 4 Permectional cystourethrography in a spina bifida boy. Note the narrow region at the external sphincter and the intraprostatic reflux of contrast medium.

are often found at adult age and cause renal insufficiency. This deterioration of kidney function is usually related to poor bladder function. The combination of urodynamic testing with nuclear renography is useful and practical.

Influence of bladder pressure on the proximal urinary tract

Several authors draw attention to the sinister implications of high bladder pressures occurring several times a day or of a chronic high pressure retention. George *et al*²² described the syndrome of high pressure chronic retention in men with obstructive uropathy: they had a late onset enuresis, a tense painless palpable bladder, hypertension and renal impairment associated with bilateral hydroureteronephrosis. McGuire *et al*²³ showed in a group of 42 myelodysplastic patients with a follow up of 7 years that in patients with an intravesical pressure

of more than 40 cm H₂O at the moment of urethral leakage, vesicoureteral reflux is seen in 68% of the cases and ureteral dilatation on excretory urography in 81%. Steinhardt *et al*²⁴ demonstrated by renal scanning in meningocele patients a consistent and significant decrease in glomerular filtration rate at high bladder pressures. In another 6 patients Zwiers *et al*²⁵ demonstrated by radionuclide renography an obstructed pattern when the bladder was filled up to the volume that during previous urodynamic testing resulted in an intravesical pressure of 40 cm H₂O. The same relationship between pressure and upper urinary tract function exists in patients with spinal cord injury, multiple sclerosis, and vesical decentralisation syndromes.

In a prospective study of 250 myelodysplasia patients McLorie *et al*²⁶ demonstrated that significant determinants of vesicoureteral reflux include a change in bladder shape, the presence of uninhibited contractions and the presence of dyssynergia. Renal deterioration was noted and predicted by the presence of vesicoureteral reflux, a high urethral pressure profile and dyssynergia. In another study on myelodysplastic patients McGuire²⁷ showed that the clinical presence of upper urinary tract deterioration may manifest itself after many years of normality and often as late as the mid teenage years; anticholinergic medication, frequently in conjunction with intermittent catheterisation, is of the utmost importance in the prevention of kidney function deterioration in patients presenting detrusor hypertonia. Jones *et al*²⁸ demonstrated that pelvic pressures are markedly elevated when the flow in the upper urinary tract was increased by oral waterload or furosemide and then accurately reflect any change in intravesical pressure. Under these conditions the pelvic pressures frequently exceeds 40 cm H₂O, which is the safety limit described by McGuire *et al*.²⁷

Bladder pressure related to changing circumstances

It should be stressed that pressure values obtained during a urodynamic session are not necessarily comparable to values exist-

ing throughout the day when the bladder is filled by natural kidney secretion. Styles *et al*^{29,30} found a 2 times higher effective cystometric capacity during standard cystometry than during long term monitoring, while detrusor instability was greater. High filling pressure commonly observed in men with chronic retention investigated by means of a standard cystometrogram are often absent during long term monitoring; they are probably due to the unphysiological filling rates and to the larger volumes of infused fluid. Therefore, the normal upper urinary tract observed in 30% of the patients showing high bladder pressures in the cystometrogram could signify that the high pressures are artificial. Nevertheless, Styles^{29,30} clearly demonstrated by long term bladder pressure monitoring that a significant correlation exists between frequently occurring uninhibited detrusor contractions, upper tract dilatation and impaired

glomerular filtration rates; the amplitude of the unstable detrusor contractions was not directly related to the upper tract dilatation.

Another situation where measured bladder pressures do not reflect the real bladder status is in patients who have a vesicoureteral reflux. Woodside *et al*¹¹ demonstrated that if the ureters with vesicoureteral reflux are occluded by a Fogarty catheter the measured intravesical pressures are much higher than those measured when urine can escape into the ureter and pelvis. Williams³¹ already considered reflux and diverticulum as a safety valve in children with low urinary tract obstructions, eg urethral valves. In this condition detrusor hypertonia can be neglected and this can result in operative failure after reconstructive surgery. Therefore reflux in the presence of significant detrusor hypertonia should be treated firstly by anticholinergic medication before performing a ureteroneocystostomy.

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