Pediatr Nephrol (2008) 23:537–540
DOI 10.1007/s00467-008-0768-3

EDITORIAL

The neurogenic bladder: introducing four contributions

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Published online: 16 February 2008

Abstract Neurogenic bladder dysfunction in children is frequently seen in patients with meningomyelocele (MMC). The disorder carries a high risk for all kinds of complications, with renal damage being the most important. More than 95% of MMC patients have a neurogenic bladder, the paramount manifestation of which is a disturbed coordination between detrusor and sphincter muscles. This vesicourethral dysfunction leads to defective filling and emptying of the urinary bladder. Voiding at will is almost never possible. According to the location and extent of the neural tube lesion, patients have either an atonic or a hypertonic pelvic floor and either an atonic or a hypertonic detrusor, leading to four classic combinations. Hypertonic sphincter and detrusor hyperactivity lead to the most dangerous form of neurogenic bladder, referred to as the "unsafe" bladder. The presence of residual urine in a high-pressure container causes either decompensation of the detrusor with vesicoureteral reflux or deterioration of the bladder wall with hypertrophy and stiffness resulting in uterovesical obstruction. The subsequent insufficient drainage of the upper urinary tract leads to decompensation of the ureters and finally to chronic renal disease, the process being accelerated by urinary tract infections. The aim of treatment is to restore as much as possible both essential functions: urine storage and timely emptying of the reservoir. What should and can be achieved is a more or less adequate, low-pressure, functional capacity of the bladder that is emptied as completely as possible by clean intermittent catheterization (CIC). MMC leads to the prototype of neurogenic bladder in childhood. What we know and what we do for MMC patients can roughly be applied to all other forms of neurogenic bladder, either congenital or acquired.

Keywords Neurogenic bladder · Neuropathic bladder · Meningomyelocele · Medical education

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The aim of the teaching series

The concern that in many centers around the world pediatric nephrologists are not sufficiently involved in the care of children and adolescents with neurogenic bladder dysfunction led to the present set of teaching papers. This concern applies particularly to the most representative group of patients: those with congenital neural tube defects. They are mainly in the care of surgical specialists for obvious practical reasons. Take, for example, a newborn with MMC, the single-most common cause of neurogenic bladder. In the first days of life, this baby will undergo closure of its open back, followed in many cases by a shunting procedure to alleviate hydrocephalus, with both procedures being carried out, supervised, and followed up by the neurosurgeon. The next step in modern management is urodynamic investigation, a technique mainly performed and interpreted by the pediatric urologist, who decides upon treatment, imaging, and follow-up. When in case of a hypertonic low-capacity bladder, medical treatment is judged insufficient, or catheterisation is too difficult, the urologist will proceed to bladder augmentation with or without a continent stoma. The third physician involved is the orthopedic surgeon when limb or spine deformities become a matter of concern. It is also no more than fair to acknowledge the decisive role of surgeons in the develop-

ment of techniques that have improved considerably not only the life expectancy of MMC patients but also their quality of life.

In many cases and places, the pediatric nephrologist is called upon in a much later state when renal damage is documented. This actually should no longer be the case. One notable exception to this rule is Jan van Gool, to whom I pay tribute here for his remarkable contributions as a pediatric nephrologist to our knowledge of the neurogenic bladder [1–3]. The upcoming series of papers on the neurogenic bladder will hopefully contribute to the awareness that the optimal approach to the patient with different forms of neurogenic bladder consists of a multidisciplinary team to surround the patient and the parents from the early stages of diagnosis and treatment. The team should include a pediatric nephrologist, besides the above-mentioned surgical specialists, as well as pediatric neurologist(s), physiotherapist(s), social worker(s), and specialized pediatric nurse(s). The role of pediatric nephrologists is obvious: they know exactly how to treat and prevent urinary tract infections, they are the experts in adequate assessment of renal anatomy and function, and they are skilled in blood pressure problems and water and electrolyte disturbances. The most important contribution of pediatric nephrologists to the team might be to strive for a balanced approach by optimizing the conservative measures and hence defining the most appropriate timing for surgery.

The four papers

It might be a surprise that none of the forthcoming papers on the neurogenic bladder has been written by pediatric nephrologists. In fact, this turned out to be no disadvantage whatsoever. I am pleased to announce that all four papers written by eminent experts in the field are of excellent quality and extremely useful for the purpose we had in mind.

Stuart Bauer, the world-famous professor of urology from Harvard Medical School, provides an in-depth review of the etiologies of the neurogenic bladder and elegantly describes how to assess these patients in a most modern fashion [4]. He reminds us that MMC is only one of the many causes of neurogenic bladder dysfunction in childhood by placing emphasis on pitfalls of occult spinal dysraphism, the exceptional but fascinating genetic forms—in particular, Currarrino syndrome with autosomal dominant inheritance—and especially the frequency of bladder problems in children with cerebral palsy. He describes in detail the subtleties of the urodynamic investigation, which is the mainstream investigation to allow an optimal, individualized approach adapted to the special needs of every patient.

Carla Verpoorten and Gunnar Buyse, two pediatric neurologists who coordinate a multidisciplinary outpatient clinic for more than 250 spina bifida children in the university hospital of Leuven, Belgium, describe in detail the principles and practice of the medical treatment for neurogenic bladder [5]. They teach us in clear terms how to identify the "unsafe bladder" that, if left untreated, leads to progressive bladder deterioration and eventual kidney damage. That is why they put so much emphasis on the presymptomatic treatment to prevent the above-mentioned and well-known gloomy scenario. On the other hand, their experience provides evidence for the reversibility of both the stiff bladder wall and vesicoureteral reflux when applying oxybutynin intravesically, along with CIC.

de Jong et al. from Utrecht, the Netherlands, present a literature overview together with a most impressive summary of their own experience with surgical aspects of neurogenic bladder management [6]. In fact, while being pediatric urologists in the first place, they apply an integrated strategy in which there is a fair place for all conservative measures as well. In their paper on the surgical aspects, they insist on early initiation of both CIC and anticholinergic drugs but also on early surgery when necessary. Their special interest and experience encompasses two procedures: the sling suspension technique for incontinence in cases of paralyzed pelvic floor; and the bladder autoaugmentation for small, high-pressure bladders. Both techniques are remarkably successful in their hands. They also highlight, as a novelty, the surgical approach to improve sexual satisfaction in young male adults with MMC.

I am most grateful to **Christopher Woodhouse**, an eminent and experienced London urologist for his astonishing contribution on the neglected aspects of MMC patients [7]. If there is general agreement that the improved management of MMC patients over the past 50 years is one of the great achievements of modern medicine, Mr. Woodhouse damps down our optimism as to the fate of young adults with congenital forms of neurogenic bladder and associated anomalies. As in many chronic conditions, the special needs of adolescents and young adults are largely either neglected or underestimated by pediatricians and pediatric surgeons. In addition, adult urologists at the time they take the patients in charge erroneously believe that these aspects have been dealt with. Mr. Woodhouse's paper, with the scarce data he was able to collect along with his personal experience in the UK, shows a dismal picture of the former pediatric MMC patient once she or he becomes an adult. Both men and women are faced with an overall poor general health condition, as reflected in a high mortality rate from cardiovascular disorders; lack of occupational, professional, and academic skills; unsatisfactory social integration; and a lot of unanswered questions

and concerns about sexuality and fertility. Mr. Woodhouse's paper is both confrontational and stimulating for all involved specialists. It voices an urgent call for increased awareness of the need to actively prepare the pediatric MMC patient for a decent quality of life in adulthood.

The global strategy

Taking together the contents and the messages formulated in these four papers, the quintessence of up-to-date management of the child with neurogenic bladder dysfunction emerges. For infants identified as belonging to the high-risk category, CIC is the central procedure to teach the parents—the earlier the better—associated with the use of anticholinergic drugs to normalize intravesical pressure. When oral intake yields insufficient results or disturbing side-effect, the intravesical route of administration should be applied. Only when this strategy fails should one consider surgery, but only after having scrutinized whether all measures prescribed were adequate and appropriately applied.

Treatment, both medical and surgical, should be adapted to the individual patient's needs. The urodynamic investigation has a central role to play in evaluating the patient and the treatment. The ultimate goal is to provide continence, to prevent bladder-wall destruction, and to preserve upper tract and kidneys function. It is not always possible to reach this fourfold goal. Therefore, one should keep in mind some priority rules. First, renal function preservation is more important than continence. Second, noninvasive measures take priority over surgery, as the former are reversible but the latter is not. This applies particularly to classic bladder augmentation. It can bring much relief but also cause serious side effects, including abundant slime production, repeated urinary tract infections, urolithiasis, stoma obstruction, and eventually tumors arising from the built-in intestinal loop. Careful weighing of the pros and cons requires input of all team members and thorough information to the parents and, as soon as she or he is old enough, the patient.

A few notes of caution

There are a few other caveats that might not be evident from reading the four contributions. A first note of caution concerns the urodynamic investigation. First, it is by no means a simple investigation, and its interpretation is not always straightforward. It has many pitfalls that must be taken into account. It should be carried out with great care by experienced personnel and whenever possible in the presence of the urologist in charge of the patient. Equally important is the collaboration of the patient, because there is a serious risk for artifacts. It is of utmost importance to document whether or not the patient is on or off any medication that interferes with detrusor and/or sphincter function. Another aspect is that urodynamics can register changes over time independently from management: one of the most important causes not to be missed is occurrence or recurrence of tethering. In this respect, the phenomenon of occult tethering deserves mentioning here [8].

Another warning is about the overdue enthusiasm for some newly propagated forms of treatment in the absence of adequate validation. This applies especially to intravesical application of the botulinum toxin as a technique to decrease detrusor hyperactivity [9]. One should not underestimate the possible side effects of this toxin, which is one of the most powerful poisons known by nature. Botulinum toxin is an exotoxin produced by spores and cells of *Clostridium botulinum*. The toxin binds to presynaptic terminals of the central nervous system and blocks the release of acetylcholine, resulting in paralysis. The doses used for local injection in the detrusor are strong enough to kill an adult human. The lethal dose of this toxin in humans is not exactly known but is derived from experiments in primates; 100 ng given i.v. or i.m., would kill a 70-kg adult. Besides its toxicity, it is good to remember that even if it is effective in a given patient—which is not always the case—it yields a short-lived result and needs to be repeated after some months.

Along the same lines, it should be stated clearly that most measures and procedures generally accepted and applied in this field are not evidence-based. In other words, there is a need for prospective and controlled studies here as in so many other fields and disorders of interest for pediatric nephrologists. Moreover, there is need for creative initiatives to solve some still unsolved problems in individual patients. As an example, I refer to a recent paper by Koff et al. who demonstrated that nighttime bladder drainage might be a simple additional measure to improve upper urinary tract deterioration in MMC patients with polyuria [10].

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