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Place of perineal electrophysiologic testing in multiple sclerosis patients

Le rôle des explorations électrophysiologiques périnéales chez des patients atteints de sclérose en plaques

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Abstract

At present, apart from visual evoked potential testing, clinical neurophysiological testing is rarely used in multiple sclerosis (MS) patients at all. In MS patients with lower urinary tract, bowel and/or sexual dysfunction, which are highly prevalent in MS, clinical neurophysiological tests of the sacral nervous system have been introduced in the past and reported as abnormal. Their usefulness is – in selected patients – to substantiate the suspicion of the neurogenic nature of the pelvic organ dysfunction, but there is no specific recommendation for MS patients. Concentric needle EMG of the perineal muscles, bulbocavernosus reflex latency measurement and pudendal SEP may be particularly useful in those rare patients in whom MS may affect the conus medullaris, to demonstrate ''directly'' this lesion of the lower sacral segments. Clinical neurophysiological testing retains the potential to further clarify interesting research questions of correlation of nervous system function and pelvic organ dysfunction, particularly relevant for application of sophisticated new rehabilitative methods including electrical stimulation of the nervous system.

Keywords: Bulbocavernosus reflex; Conus medullaris; Multiple sclerosis; Neurophysiological tests; Sacral nervous system

Résumé

En dehors de l'examen des potentiels évoqués visuels, les explorations électrophysiologiques n'ont guère de place chez les patients atteints de sclérose en plaques (SEP). Chez les patients qui souffrent de dysfonction vésico-sphinctérienne et/ou sexuelle, dont la prévalence dans cette pathologie est importante, les examens neurophysiologiques du système nerveux au niveau sacré ont été introduits par le passé et les résultats ont été considérés comme anormaux. Leur utilité dans certaines populations consiste à étayer le soupçon de la nature neurogénique de la dysfonction pelvienne, mais il n'existe pas de recommandation spécifique pour les patients atteints de SEP. L'EMG avec aiguille concentrique des muscles périnéaux, l'évaluation de la latence du réflexe bulbocaverneux et l'enregistrement du potentiel évoqué somato-sensoriel (PES) pudendal peuvent être d'une grande utilité chez les rares patients dont la SEP touche le cône terminal comme moyen de démontrer « directement » la présence de cette lésion des segments médullaires inférieurs. Des examens de neurophysiologie clinique pourraient permettre d'éclaircir des points spécifiques de recherche sur la corrélation du fonctionnement du système nerveux et la dysfonction pelvienne et éventuellement contribuer à l'application de nouvelles méthodes rééducatives sophistiquées, dont la stimulation électrique du système nerveux.

Mots clés : Réflexe bulbocaverneux ; Cône terminal ; Sclérose en plaques ; Examens neurophysiologiques ; Système nerveux au niveau sacré

1. English version

1.1. Introduction

Multiple sclerosis (MS) is a chronic, autoimmune, inflammatory, demyelinating, neurodegenerative disease of the central nervous system. It is the commonest progressive neurological disorder in young adults, affecting more than 2 million people worldwide [1]. Women are affected twice as often as men (ratio from 1.1 to 3.0 across different regions) [2]. The prevalence shows a high variability across regions and populations. According to its clinical course, four subtypes have been identified. Relapsing remitting MS (RRMS) is the commonest (85% of patients), but over a median period of 11

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years nearly half of these patients convert to secondary progressive MS (SPMS) [3]. About 10% of patients have progressive symptoms from the onset (primary progressive MS, PPMS) and a minority have progressive relapsing MS (PRMS) [4]. The disorder almost invariably progresses, leading to increasing disability.

The major histopathologic changes in MS consist of the loss of myelin and axonal damage due to inflammatory and neurodegenerative mechanisms [5]. Relapsing remitting MS is characterized by new and active focal inflammatory demyelinating lesions in the white matter, while diffuse injury of normal appearing white matter, cortical demyelination and axonal loss are characteristic of primary and secondary progressive multiple sclerosis [6].

Common clinical manifestations of MS include spasticity in 40–85%, fatigue in 69–97%, ataxia and tremor in up to 80%, pain (acute or chronic; neuropathic, somatic or psychogenic) in 29–86%, optic neuritis in up to 70%, and cognitive impairment in 40–70% of patients during the course of the disease [7]. However, bladder dysfunction (in 80–96%) and bowel dysfunction (constipation and/or incontinence in 29–54%) are also common [7]. Dysfunctions of pelvic organs in patients are, as can be seen in practice, aggravated by other MS symptoms, including spasticity, pain, fatigue, cognitive deficit etc.

Estimates of the proportion of patients with MS who have lower urinary tract (LUT) symptoms vary according to the severity of the neurological disability in the group under study, but a figure of about 75% is frequently cited [8]. However, it seems that when more detailed investigation protocol is used, including urodynamics (cystometry and pressure-flow study) and uro-neurophysiological investigations (sacral reflex studies and pudendal SEP), majority (88%) of patients demonstrate abnormalities even in the early disease phase (e.g., mean disease duration 5 years) [9]. Cystometric studies in groups of patients with MS have found detrusor overactivity (DO) as the most frequent abnormality (mean occurrence 65%) followed by detrusor underactivity (mean occurrence 25%) and poor bladder compliance (2% to 10%). The prevalence of detrusor-sphincter dyssynergia (DSD) varied in different studies (median prevalence of 35%), increasing with disease progression over time [10].

Several studies have shown that urinary incontinence is considered to be one of the worst aspects of the disease; with 70% of a self-selected group of patients with MS responding to a questionnaire classified the impact of bladder symptoms on their life as ‘‘high’’ or ‘‘moderate’’ [11]. Symptoms, particularly urgency, incontinence and nocturia, can limit activities of daily living of MS patients. They also negatively impact on a patient’s social activities, leading to social embarrassment, isolation and depression [12].

Considering the multitude of symptoms reported by MS patients, unsurprisingly, LUT symptoms may be over-looked. The North American Research Committee On Multiple Sclerosis questionnaire survey in more than five thousand MS patients with troublesome urinary symptoms revealed that only 43% had been referred to urological services and 51% had been treated with antimuscarinic medications [13]. Recently, a screening tool for patients with bladder problems related to MS called the ‘Actionable Bladder Symptom Screening Tool’ has been developed and validated [14].

Sexual dysfunctions (SD) are also highly prevalent in MS patients and include diminished desire, arousal/erectile dysfunction and orgasmic/ejaculatory dysfunction. SD’s are reported in 64 to 91% of MS men, most commonly ED (19–62%) [15]. Other frequent complaints include decreased sexual desire, decreased sensation during sexual stimulation, ejaculatory and orgasm dysfunction [15]. Significant differences are found between authors in the estimation of decrease in sexual desire (31–64%) and impaired arousal (33–52%) and much less when it came to evaluating orgasmic difficulties (37–38%) in MS women [16].

In reviews on MS, it is as a rule asserted that pelvic organ dysfunction is mainly due to spinal cord disease, and a correlation between LUT symptoms and the degree of pyramidal symptoms/signs in the lower limbs was reported [17]. Nevertheless, LUT, bowel and SD in MS can obviously be caused by damage to the brain, spinal cord, and to the peripheral neurons engaged in the organs’ neural control.

1.2. The need for assessment of lower urinary tract (LUT) function in multiple sclerosis (MS) patients

A UK consensus paper recommended that urodynamics should be carried out only in those MS patients who are refractory to conservative treatment or bothered by their symptoms and wish to undergo further interventions [17]. Similar recommendations have been put forward by the NICE recommendations for urinary incontinence in neurological disease [18]. By contrast, the French authors advocate use of urodynamic testing generally in MS bladder symptoms management planning [19].

1.3. Clinical neurophysiological assessment in multiple sclerosis (MS) patients

According to revised McDonald criteria, MS diagnosis is nowadays made on basis of clinical features (two or more relapses), neuroimaging findings (two or more lesions consistent with MS), and cerebrospinal fluid examination (intrathecal synthesis of IgG or at least two oligoclonal bands) [20]. After introduction of MRI studies in MS diagnosis, neurophysiological studies became much less important. Currently, in most places, only visual evoked potentials are used selectively to supplement MRI findings in demonstration of dissemination of demyelinating lesions in space, other SEP studies being performed, rarely, if at all. Evoked potentials, however, are occasionally useful in MS diagnosis, because they can better discriminate between demyelinating and axonal lesions than MRI; small vascular and demyelinative lesions may look very similar.

Sacral electrodiagnostic studies are as a rule not used to diagnose MS. However, rarely MS may present with uro-ano-genital dysfunction, most commonly caused by a conus