

KEYPOINTS:

- Although genito-urinary tract abnormalities represent some of the most demoralizing and disabling manifestations of MS, there continues to be a lamentable code of silence between physicians and patients concerning these important issues.
- Abnormalities of the lower urinary tract (bladder and urethra) will be experienced by at least 96% of MS patients after 10 years of disease activity.

CHAPTER 5

MULTIPLE SCLEROSIS (MS), OTHER MYELOPATHIES AND SPINAL CORD INJURY (SCI)

INTRODUCTION

A comprehensive assessment and treatment strategy for bowel, bladder, and sexual dysfunction in a patient with multiple sclerosis (MS), other myelopathies, or spinal cord injury (SCI) requires a co-ordinated approach based on a knowledge of the patient’s disease course, iatrogenic factors, concomitant symptomatic complaints, as well as a knowledge of their social and psychological factors. A multidisciplinary team is often required involving neurologists, urologists, gynecologists, rehabilitation specialists, nurses, and primary care physicians. Through their efforts, the function and quality of the patient’s life can be significantly improved.

MULTIPLE SCLEROSIS (MS)

Although genito-urinary tract abnormalities represent some of the most demoralizing and disabling manifestations of MS, there continues to be a lamentable code of silence between physicians and patients concerning these important issues. Without exception, patients should be asked about bladder, bowel, and sexual function. A comprehensive exploration into the problems can facilitate the diagnosis and treatment of such ailments. A greater understanding of the pathophysiology, diagnostic techniques, and treatment interventions for genitourinary dysfunction will equip the neurologist to manage patients with MS more effectively.

BLADDER DYSFUNCTION

A set of questions should be used to elicit information from the patient (Table 1). Furthermore, an interdisciplinary strategy of assessment and intervention that emphasizes the collaboration between the neurologist and urologist optimizes the broadest range of treatment

options and enhances the chances for successful management of the MS patient with genitourinary tract dysfunction.

Abnormalities of the lower urinary tract (bladder and urethra) will be experienced by at least 96% of MS patients after 10 years of disease activity. Bladder dysfunction can produce a diversity of symptomatic complaints that are differentiated on the basis of whether they relate to storage or emptying deficits. The most common complaints relating to bladder dysfunction in patients with MS are urgency, frequency, and urge incontinence, all of which are related to detrusor hyperreflexia. Others include hesitancy, interrupted stream, incomplete emptying, double voiding, nocturia, and enuresis.

While MRI imaging has substantially advanced our ability to diagnose patients with MS and follow disease activity, there has been no recognized correlation between the number of lesions or atrophy and any specific urodynamic parameter or bladder dysfunction [1].

TABLE 1 Question set for evaluating bladder dysfunction

1. Frequency
2. Urgency
3. Urge incontinence
4. Frank incontinence
5. Stress incontinence
6. History of obstetrical injury or genitourinary surgery
7. Hesitancy
8. Double voiding
9. Nocturia and/or enuresis
10. Iatrogenic agents

However, a number of features of the clinical examination appear to correlate with lower urinary tract dysfunction and lower extremity motor dysfunction seems to be the best predictor of bladder dysfunction. In a study with 170 MS patients and bladder symptoms, only four did not have symptoms and signs of pyramidal abnormalities [2]. Extensor plantar responses (Babinski's sign) are evident in 70-95% of patients with bladder dysfunction and in 70% with detrusor sphincter dyssynergia (DSD). This strong positive relationship between bladder abnormalities and pyramidal dysfunction in the lower extremities is not surprising considering the neuroanatomical juxtaposition of these distinctive pathways, with the 'legs above the bladder' (Figure 1). Other features include hyperreflexia, clonus, spasticity, pyramidal distribution weakness, and a sensory level (all suggestive of a myelopathy). There is also a correlation between urinary symptoms and lesions with the midbrain and pons [3-5]. In particular, specific brainstem syndromes are associated with a higher risk of bladder dysfunction, especially DSD. The most notable of these syndromes is internuclear ophthalmoplegia, a neuro-ophthalmologic hallmark of MS.

While detrusor hyperreflexia with synergic voiding is the most common pattern of bladder dysfunction in MS, the incidence of DSD has been estimated to range from 18 to 66%. The uncoordinated dyssynergia is believed to result from an interruption in the descending regulatory pathways from the pontine micturition center such that there is simultaneous contraction of the detrusor

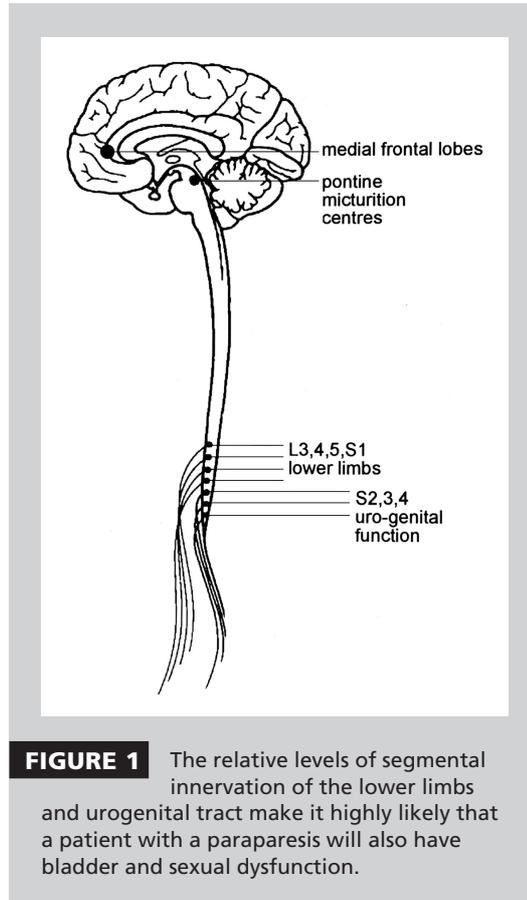


FIGURE 1 The relative levels of segmental innervation of the lower limbs and urogenital tract make it highly likely that a patient with a paraparesis will also have bladder and sexual dysfunction.

muscles and the urethral sphincters. The consequences of DSD can include a high post voiding residual (PVR), increased bladder pressures, a higher risk of urinary infections and urinary tract calculi, and upper tract disease — principally hydronephrosis, in occasional patients.

KEYPOINTS:

- Strong positive relationship between bladder abnormalities and pyramidal dysfunction in the lower extremities is not surprising considering the neuroanatomical juxtaposition of these distinctive pathways, with the 'legs above the bladder'.

CASE 1: BLADDER DYSFUNCTION

ML is a 41-year-old female patient with a 5-year history of relapsing-remitting MS. She has evidence of diffuse hyperreflexia, mild lower extremity spasticity, bilateral extensor plantar responses and subtle but definite bilateral internuclear ophthalmoplegia. She complains of frequency, hesitancy, urgency, and urge incontinence with two-time nocturia.

Treatment considerations

1. Measure a PVR, urinalysis, urine culture and sensitivity, BUN, creatinine, blood glucose, renal ultrasound.
2. If the PVR is less than 100 ml, treat with anticholinergic agent and repeat the PVR in 1-2 weeks.
3. If the PVR is greater than 100 ml and not reduced with double voiding or the application of a hand-held vibrator applied to the lower abdomen, begin intermittent catheterization and add anticholinergic agents for urgent bladder symptoms. Consider referral to a urologist for urodynamics.
4. If nocturia is not improved with the above measures, consider adding nighttime DDAVP.

NEUROLOGIC BLADDER, BOWEL AND SEXUAL DYSFUNCTION

KEYPOINTS:

- It is important not to rely on the history in order to ascertain whether voiding dysfunction is present. For instance, in one study, only 47% of patients with an elevated PVR had a sensation of incomplete emptying [2]. However, of those who claimed to empty their bladders completely, 53% were incorrect.
- For the majority of patients with MS that a general neurologist will evaluate, a PVR is often the only assessment that is necessary.
- The emergence of effective treatment strategies for sexual dysfunction provides compelling justification to overcome the reluctance to explore these issues in a sensitive and objective manner.
- Estimates suggest that the incidence of sexual dysfunction in MS ranges from 62 to 83%.

In a smaller percentage of patients, detrusor hypocontractility and areflexia can occur in association with large PVRs. This pattern may result from loss of cortical facilitation, from cerebellar lesions, or from lesions below the sacral cord.

It is important not to rely on the history in order to ascertain whether voiding dysfunction is present. For instance, in one study, only 47% of patients with an elevated PVR had a sensation of incomplete emptying [2]. However, of those who claimed to empty their bladders completely, 53% were incorrect [2]. Because of the effect of a raised post void residual volume on bladder function it is essential to measure the PVR before prescribing anticholinergic medication, as emphasized in Chapter 2.

Upper tract disease is exceptional in MS and is linked to predictable risk factors including DSD and the presence of an indwelling catheter. This is in marked contrast to those with spinal cord injury who are very much at risk of developing upper urinary tract complications. The reason for this difference is not known since MS can be associated with just as severe lower limb spasticity as may occur following SCI.

For the majority of patients with MS that a general neurologist will evaluate, a PVR is often the only assessment that is necessary. However, in selected patients, urodynamic evaluation, as discussed in Chapter 2, can facilitate the proper identification of bladder and sphincteric abnormalities. This assessment can also provide compelling evidence to suggest upper motor neuron dysfunction in patients with a suspected CNS process such as MS. When no formal urinary investigation is completed, 73% of MS patients are treated inappropriately [6]. At a minimum, a PVR should be completed in all patients with MS on a periodic basis in order to capture evidence of voiding dysfunction.

SEXUAL DYSFUNCTION

While sexual abnormalities represent some of the most distressing features of MS, they are often not discussed by patients with their physicians during the process of medical care. In a study involving 47 women with MS and sexual dysfunction, the majority had never

TABLE 2 Question set for evaluation of sexual dysfunction

1. Sexually active either with partner or by self stimulation
2. Communication with sexual partner
3. Interest in sex (libido)
4. Arousal
5. Sensation
6. Ability to achieve orgasm
7. Erection
8. Lubrication
9. Pain
10. Fatigue
11. Sexual practices and positions
12. Spasticity and its impact
13. Bladder dysfunction during sex

been asked to discuss their sexuality in relation to their disease [7].

The emergence of effective treatment strategies for sexual dysfunction provides compelling justification to overcome the reluctance to explore these issues in a sensitive and objective manner. A comprehensive assessment of the patient's various symptomatic complaints is imperative in order to maximize sexual functioning (Table 2).

Chapter 4 discusses in general the various options for treating neurogenic sexual dysfunction. Specifically in MS, the therapy for fatigue, spasticity, pain, bowel and bladder dysfunction, and mood disorders should be optimized, as these various factors can directly impair sexual functioning. Even if therapeutic strategies are not effective in individual patients, the process of open and honest exploration of these issues can be an important and meaningful experience for both the patient and physician. A number of myths and misconceptions often serve as a powerful obstacle to pursuing education and treatment of sexual disorders (Table 3).

Estimates suggest that the incidence of sexual dysfunction in MS ranges from 62 to 83% [8-10]. Concomitant conditions such as diabetes and atherosclerosis can more strongly predispose patients to develop a sexual disturbance.

TABLE 3 Myths and misconceptions of sexual dysfunction in MS

1. I have MS and sexual dysfunction is the least of my concerns
2. Disabled patients have no sexual needs
3. My physician is not interested in hearing about sexual dysfunction
4. It is not appropriate to discuss sexual dysfunction with my doctor
5. Sex must culminate in orgasm
6. Sex must involve intercourse
7. Physical contact must involve sexual activity
8. People should instinctively be sexually competent
9. Patients should not masturbate or use fantasy

Sexual dysfunction in men with MS. Patients with MS will commonly experience sexual dysfunction, often in combination with urinary abnormalities. The initial assessment should include a search for factors that potentially contribute to sexual abnormalities (Table 4). Erectile dysfunction (ED) will occur in about 70% of men with MS [11-13]. The mean time between the onset of neurological symptoms and the emergence of ED in MS patients is about 9 years: sexual dysfunction is rarely a presenting manifestation of MS [13,14]. As with

patients with MS and urinary dysfunction, there is a strong correspondence between ED and pyramidal dysfunction in the lower extremities. Although there has been an interest in the relationship between sexual dysfunction in MS and neuroendocrine changes such as testosterone deficiency, a correlation has not been confirmed.

Men with MS may still experience spontaneous erections, particularly at night i.e. nocturnal penile tumescence, but the presence of nocturnal erections does not indicate that their problem is psychiatric, as was perhaps formerly thought. The ability to sustain an erection adequately for intercourse depends largely on psychogenic erectile drive which is impaired by spinal cord disease.

A variety of neurophysiological tests have been used to assess ED in MS, including the pudendal evoked potential (see Chapters 1 and 4). However, the current view is that these do not contribute to understanding the complaint of ED in a man with MS who on clinical examination has evidence of spinal cord disease.

KEYPOINTS:

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- Men with MS may still experience spontaneous erections, particularly at night i.e. nocturnal penile tumescence but the presence of nocturnal erections does not indicate that their problem is psychiatric, as was perhaps formerly thought.

CASE 2: MALE SEXUAL DYSFUNCTION IN MS

SL is a 35-year-old man with clinically definite secondary progressive MS with ongoing relapses. His clinical course has been stabilized with interferon therapy. He has mild spastic paraparesis and complains of sexual dysfunction characterized by decreased ability to achieve and maintain erection, difficulty achieving orgasm, chronic fatigue, and frequent extensor and adductor spasms during sexual intercourse. His wife is concerned that her husband no longer finds her attractive. Their primary sexual activity has consisted of intercourse.

Treatment considerations

1. Begin a broad discussion about sexual dysfunction in MS with the patient and his wife.
2. Encourage more communication between the husband and wife.
3. Assess for any iatrogenic causes.
4. Optimize treatment for spasticity, especially before sexual activity.
5. Evaluate sleep hygiene and treat nocturnal spasms and nocturia if present.
6. If there are no contraindications, consider treating erectile dysfunction with sildenafil.
7. If sildenafil is ineffective, consider alternative treatment, such as intracorporeal alprostadil.
8. If orgasm is still difficult to achieve, suggest that vibrator stimulation be applied to the ventral surface of the glans penis (at the frenulum).
9. Emphasize the value of energy-conserving sexual positions and encourage alternative sexual practices other than intercourse.

NEUROLOGIC BLADDER, BOWEL AND SEXUAL DYSFUNCTION

TABLE 4 Factors that influence sexual dysfunction in MS and SCI

1. Dysfunction can fluctuate with disease activity
2. Reduced sensory responsiveness
3. Psychological factors and depression
4. Pain and paresthesias
5. Bowel and bladder dysfunction
6. Spasticity
7. Fatigue
8. Iatrogenic causes (e.g. SSRIs, tricyclic antidepressants)
9. Relationship discord
10. Physician neglect ('code of silence')
11. Preoccupied with misconceptions
12. Alteration of body image

Oral therapy for MED in MS. While a variety of therapeutic options are currently available for men with ED, oral agents have clearly eclipsed all other therapies in terms of popularity and effectiveness. Along this line, the most significant advance has been the development of oral sildenafil citrate (*Viagra*®) (see Chapter 4). The response of men with MS to treatment with this agent has been excellent [15]: comparisons across

studies showing a particular efficacy in this patient group. Ejaculatory difficulties may, however, persist.

Sexual dysfunction in women with MS. Women with MS will frequently experience sexual dysfunction, either related to the disease process or secondary to other factors that similarly affect males (Table 4). Diminished lubrication can be improved by the liberal use of lubricating agents that are now widely available. Some women will experience sensory disturbances during sexual activity that can often be treated effectively with membrane stabilizing medication such as gabapentin, carbamazepine, and similar agents.

In those who experience vaginal hypesthesia, most often secondary to sensory myelitis, vibratory stimulation of the clitoris and vaginal tissues can lead to pleasurable sensations and even orgasm.

Given the similarities between male and female sexual responses, there is rationale to study the potential role of agents such as sildenafil in the treatment of female sexual dysfunction. We are currently completing a randomized, double-blind, placebo-controlled cross-over study of sildenafil in women with MS and sexual dysfunction.

CASE 3: SEXUAL DYSFUNCTION IN A WOMAN WITH MS

GN is a 58-year-old post-menopausal woman with secondary progressive MS. She complains of lower extremity spasticity, chronic fatigue, depression, and bladder urgency. There has been difficulty with diminished sexual arousal, numbness in the vaginal region, poor lubrication, painful sensations on attempted intercourse, and difficulty achieving orgasm. During sexual activity she experiences involuntary leg spasms with associated bladder urgency and urge incontinence. Her depression is well treated with fluoxetine.

Treatment considerations

1. Strongly consider discontinuation of fluoxetine and treating her depression with an alternative agent that is less associated with sexual dysfunction (e.g. trazodone, venlafaxine).
2. Assess sleep hygiene and treat nocturnal motor spasms and bladder dysfunction if present.
3. Optimally treat spasticity.
4. Treat bladder urgency and urge incontinence, especially before sexual activity (void before sex and use anticholinergic agents for bladder relaxation).
5. Encourage the use of lubricating agents.
6. Treat painful sensations (consider a tricyclic antidepressant, which may be helpful for pain, depression, and bladder urgency).
7. Refer to gynecologist for possible hormone replacement therapy.
8. Encourage energy-conserving positions.
9. Emphasize the benefits of good communication with partner concerning problems and potential solutions.

BOWEL DYSFUNCTION

Bowel dysfunction occurs in many patients with MS at some point during the course of the illness and can persist as a chronic manifestation of the disease [16,17]. Although there is a predilection for bowel dysfunction to occur in those patients with bladder complaints, in a study of patients with MS attending a clinic for treatment of urinary symptoms, one third did not have bowel problems [17].

A number of factors contribute to the onset of bowel dysfunction that must be recognized by the treating physician so that a strategic therapeutic plan can be formulated. The assessment of bowel abnormalities must begin with an inquiry about bowel symptoms and complaints (Table 5).

The most frequent abnormalities of bowel dysfunction are constipation, impaction, and incontinence, but by far the most common problem is constipation. Unlike the high frequency of bladder incontinence that occurs in MS patients, regular bowel incontinence is less common.

Constipation. The contributing factors that result in constipation are manifold (Table 6). A reduction in neuro-gastrointestinal signaling through the autonomic nervous system has been advanced as a hypothesis on how MS can lead to a slowing in intestinal transit time [18].

MS patients can experience difficulty with voluntary transitions from the storage to defecation process in response to a full rectum. Inadequate relaxation of the pubococcygeus and sphincter muscles can prohibit defecation despite the patient's perception of bowel urgency and the performance of the Valsalva

maneuver. Abdominal muscle contractions facilitate the process of fecal evacuation. These muscles are innervated by T6-T12 spinal cord segments and disease activity within the spinal cord at these levels can lead to an increase in abdominal compliance.

Following the ingestion of a meal, especially in the morning, many individuals experience the perceived need to defecate. This process is recognized as the gastrocolic reflex, but it may be blunted in patients with MS. Nevertheless, patients should be encouraged to identify the time of day where the post-meal urge to defecate is strongest.

Inadequate fluid hydration is a major contributor to bowel constipation in MS patients. Many patients recognize that fluid intake can provoke bladder urgency and urge incontinence and will therefore restrict their fluid intake. It is crucial that an integrated treatment plan be formulated for each patient that takes into consideration the interaction between bowel and bladder pathophysiological mechanisms.

Other factors that contribute to constipation include iatrogenic causes, primarily from medications, inadequate dietary or supplemental fiber, immobility, and inadequate exercise. The most common agents that can cause or exacerbate constipation in MS patients include antihypertensive agents, anticholinergics (commonly used for bladder urgency), sedative-hypnotic agents, calcium supplements and aluminum containing antacids, tricyclic antidepressants, narcotic analgesics, diuretics, and iron supplements.

When constipation is prolonged, bowel incontinence or impaction can ensue. Treatment of impaction involves the use of hyperosmotic agents, enemas, and typically disimpaction. During this time, bulking agents should be discontinued until the colon is evacuated.

TABLE 5 Question set for evaluating bowel dysfunction

1. Problems with constipation
2. Problems with diarrhea
3. Alternating bouts of constipation and diarrhea
4. Problems with incontinence
5. Bowel urgency
6. Urgency with difficulty evacuating rectum
7. Hard painful stools
8. Adequate hydration and fiber
9. Iatrogenic agents

KEYPOINTS:

- A number of factors contribute to the onset of bowel dysfunction that must be recognized by the treating physician so that a strategic therapeutic plan can be formulated.
- The most frequent abnormalities of bowel dysfunction are constipation, impaction, and incontinence, but by far the most common problem is constipation.

TABLE 6 Constipation: contributing factors

1. Reduced neurogastrointestinal signaling
2. Reduced gastrocolic reflex
3. Pelvic floor muscular spasticity
4. Inadequate hydration
5. Drugs (e.g. anticholinergic agents)
6. Immobility
7. Lack of physical conditioning
8. Weak abdominal muscles

NEUROLOGIC BLADDER, BOWEL AND SEXUAL DYSFUNCTION

KEYPOINTS:

- Fecal incontinence in MS is relatively uncommon and generally is associated with poorly treated constipation. Isolated bowel incontinence should be investigated separately.

More severe consequences can occur and include intestinal obstruction, intussusception, and rectocele. Chronic straining can cause intussusception and intrarectal obstruction leading to severe difficulty with rectal emptying or a constant sensation of incomplete rectal emptying and the need to defecate. Patients with such complaints should be referred to a colo-rectal specialist.

Fecal incontinence. When fecal incontinence occurs, a number of factors may be responsible and include reduced sensation of rectal filling, poor voluntary contraction of the anal sphincter and pelvic floor musculature (poor voluntary squeeze pressure), and reduced rectal compliance.

Fecal incontinence in MS is relatively uncommon and generally is associated with poorly treated constipation. Isolated bowel incontinence should be investigated separately. The institution of a formal bowel program for treatment of constipation (see Chapter 3) will generally reduce the risk of incontinence. Furthermore, it is important to identify provocative factors (e.g. irritants, such as caffeine and alcohol) that can be altered to reduce the incidence of incontinence episodes. In some patients with severe spinal cord dysfunction and loss of voluntary control, frequent episodes of fecal incontinence will mandate the use of protective pants and timed

elimination periods by the use of scheduled suppository treatment to establish a program of reliable bowel movements. The predictability of this regimen is of great benefit to both those demoralized by the experience of bowel accidents and their carers.

The general treatment of constipation and fecal incontinence is described in Chapter 3.

OTHER MYELOPATHIES INCLUDING HUMAN T-LYMPHOTROPIC VIRUS-1 ASSOCIATED MYELOPATHY (HAM)

Various causes of myelopathy are given in Table 7. The approach to pelvic organ dysfunction in myelopathic disorders that cause upper motor neuron bladder dysfunction is the same as that detailed above.

One example of a condition that produces similar genitourinary dysfunction to that of MS is HAM, also known as tropical spastic paraparesis (TSP) [19]. This condition is caused by a retrovirus which has an expanding worldwide distribution. The risk factors for transmission include i.v. drug abuse, blood transfusion, and sexual intercourse. The virus can also be transmitted vertically from mother to infant during breast-feeding. The diagnosis of TSP can be made by a number of molecular techniques including immunofluorescence, ELISA, Western blot, with CSF antibodies, and by PCR.

CASE 4: BOWEL DYSFUNCTION IN MS

JH is a 30-year-old nurse with a history of clinically definite relapsing-remitting MS. She presents for a follow-up clinic visit complaining of severe constipation. She previously would have daily bowel movements in the morning after breakfast. She now works the morning shift and often skips her morning meal. She has stopped her previous vigorous exercise routine and has gained 10 kg. It is clear that she has had some degree of abdominal distention secondary to her weight gain and poor abdominal muscle tone. The patient is well known to restrict fluids secondary to severe bladder urgency and a history of urge incontinence while at work. In addition, she is being treated with amitriptyline for depression and migraine headache prophylaxis. She has used laxatives and digital stimulation to promote bowel movements.

Treatment considerations

1. Resume a regular exercise program to include abdominal muscle exercises.
2. Begin a weight loss program.
3. Perform a PVR and if less than 100 ml implement anticholinergic therapy in order to reduce bladder urgency and urge incontinence. Subsequently repeat the PVR to exclude anticholinergic induced urinary retention.
4. Strongly emphasize the importance of adequate fluid hydration of at least 2000 ml/day.
5. Increase dietary fiber and begin a bulking agent.
6. Suggest the use of glycerin or other suppositories if pelvic floor and spincteric muscular spasticity is suspected.
7. Alternative agents for depression and migraine could be considered.

TSP is characterized pathologically by a chronic inflammatory process primarily affecting the spinal cord white matter. Evidence of demyelination with axonal sparing in association with macrophage activation and oligodendrocyte apoptosis reminiscent of the pathology in MS. However, in TSP there is an additional component of pathology within the bladder lamina propria in conjunction with thickened neurons which assume the appearance of 'sausage rolls' [19]. These histopathological features are distinctive to TSP and have not been observed in other myelopathies that produce bladder dysfunction.

Assessment and management of genitourinary dysfunction in patients with TSP is similar to that of other myelopathies, as outlined in Chapter 2.

Bladder dysfunction occurs in 75-100% of infected individuals with the most common patterns being detrusor hyperreflexia and DSD. Approximately 60% of TSP patients will require the use of CIC at some point during the illness [19]. Occasional patients will exhibit evidence of decreased detrusor activity, which would appear to result from disease involvement of the posterior and anterior nerve roots in addition to Onuf's nucleus. Similar to MS, upper tract disease is distinctly uncommon.

SPINAL CORD INJURY (SCI)

Bladder dysfunction. In those patients who experience spinal cord injury (SCI), there is an initial period of cessation of spinal reflexes including bladder areflexia. This typically lasts for days to weeks during which the bladder requires continuous drainage, either by a urethral or suprapubic catheter.

Many of the same assessment and treatment strategies outlined for myelopathies as described in Chapter 2 can be applied to those with SCI. However, there are major urological differences between those with progressive spinal cord disease and the spinal cord injured. The former are usually not suitable to undergo urological surgery, whereas those with SCI are otherwise fit and need a definitive solution to their bladder dysfunction, and furthermore, because they are at risk of renal failure, they should remain under urological supervision.

The risk of renal failure is due to three principal disorders: obstruction; vesicoure-

TABLE 7 Causes of myelopathy

- ▶ Spinal cord injury
- ▶ Compressive-disc disease/spondylosis/chiari malformation
- ▶ Metastatic lesions
- ▶ Spinal cord tumors (glioma, lymphoma, etc.)
- ▶ Multiple sclerosis (MS)
- ▶ Post infectious or post vaccinal myelitis
- ▶ Idiopathic transverse myelitis
- ▶ Neuromyelitis optica (Devic's syndrome)
- ▶ Neurosarcoidosis
- ▶ Amyotrophic lateral sclerosis (ALS)
- ▶ Primary lateral sclerosis (PLS) (bladder dysfunction exceptional)
- ▶ HIV (vacuolar myelopathy)
- ▶ HTLV-1 associated myelopathy (tropical spastic paraparesis)
- ▶ Leukodystrophy (e.g. adrenomyeloneuropathy)
- ▶ Nutritional causes (e.g. Vitamin B-12 deficiency)
- ▶ Hereditary myelopathies

teric reflux; and the formation of calculi. Urinary obstruction leads to an increase in bladder pressures and a thickened trabeculated bladder wall. Vesicoureteric reflux occurs secondary to elevated detrusor pressures and requires intervention with bladder drainage and the use of anticholinergic agents. Urinary tract calculi occur more commonly in those who are immobilized for prolonged periods. Stone formation can serve as a nidus for recurrent urinary tract infections. In those with chronic indwelling catheters, the balloon can be the site for the formation of eggshell calcification ultimately leading to sloughing and the formation of bladder stones. Treatments for calculi include lithotripsy, percutaneous nephrolithotomy, and ureterorenoscopy. Chronic catheterization can be associated with a small risk of squamous cell carcinoma that is substantially higher than that seen in the general population and because of this, periodic cystoscopy is recommended. While renal failure was previously responsible for significant amount of mortality in those with SCI, new management strategies have substantially reduced morbidity and mortality. An important review on this subject has recently been published [20].

In some individuals, implantation of a Brindley sacral root stimulator may be appro-

KEYPOINTS:

- There are major urological differences between those with progressive spinal cord disease and the spinal cord injured. The former are usually not suitable to undergo urological surgery, whereas those with SCI are otherwise fit and need a definitive solution to their bladder dysfunction, and furthermore, because they are at risk of renal failure, they should remain under urological supervision.

NEUROLOGIC BLADDER, BOWEL AND SEXUAL DYSFUNCTION

KEYPOINTS:

- Ninety-five percent of SCI patients required at least one therapeutic method to initiate defecation and half of them became dependent on others for toileting.
- Most men following SCI are able to obtain reflex erections, but these are not usually adequate for intercourse and few experience psychogenic erections.

appropriate. The procedure requires sacral deafferentation causing loss of reflex detrusor and erectile function and is therefore only indicated in those with a complete cord lesion. Stimulating electrodes are placed on the anterior roots of S2-S4 and a radioreceiver is placed in the lower anterior chest. Micturition can be initiated by the application of a mirror-image radio transmitter over the receiver. Following a defined period of stimulation the current is terminated. During this phase, the sphincter will relax rapidly, while the detrusor contraction does so more slowly. It is during this phase of dyssynergic muscle relaxation that effective voiding occurs.

A distinctive abnormality that occurs in those with SCI is *autonomic dysreflexia*. This syndrome generally occurs when the level of the insult is above T6. Patients can experience crescendo hypertension, severe headache, and increased risk of stroke. These changes appear to be related to episodic massive sympathetic discharges within the spinal cord, culminating in vasospasm of the skin and visceral organs. These episodes are typically precipitated by noxious stimuli below the level of the lesion which include constipation, fecal impaction, bladder contractions or can occur during bladder or bowel treatment procedures. Therapeutic considerations for those with autonomic hyperreflexia include elevation of the head of the bed and the use of antihypertensive agents, especially calcium channel blockers. Anesthetic agents can also be used before manipulations that may trigger these reactions.

Bowel dysfunction in SCI. A study of spinal cord injured patients showed that bowel dysfunction is a major physical and psychological problem [21]. Nausea, diarrhea, constipation and fecal incontinence were all much more common after SCI, but the major problems arise as a result of loss of voluntary control of defecation. Ninety-five percent of SCI patients required at least one therapeutic method to initiate defecation and half of them became dependent on others for toileting. The procedure was prolonged and in almost half bowel evacuation took more than 30 minutes and the time taken correlated with measures of distress and depression. On a scale of 0 (no perceived problem) to 10 (maximum perceived problem), patients rated their loss of

mobility as a mean of 6.8 (SD 3.3), sexual satisfaction 5.5 (SD 4.2), urinary management 4.9 (SD 3.5) and bowel management 5.1 (SD 3.6).

Optimizing bowel function can help to restore dignity and control to a person following SCI. Failure to avoid the complications of constipation can result in significant morbidity (e.g. bed sores, anal fissure). Furthermore, some 10% of SCI fatalities may be ascribed to undiagnosed abdominal emergencies. Management of bowel problems requires effort on the part of both patient and carer, who must set aside time regularly for bowel care.

Management. Establishment of a bowel program should be individualized. Patients will have different bowel patterns in terms of the frequency of bowel movements. It is important to recognize when an individual patient is more likely to produce stool. For some patients this will be in the morning while in other it will be later in the day. The majority of SCI patients practice regular manual evacuation to prevent impaction and reduce the risk of autonomic dysreflexia. In those SCI patients with a preserved defecation reflex, reflex emptying can be stimulated every 1-3 days by gentle rotation of a gloved finger in the anus until the rectal wall is felt to relax, or flatus is passed and the stool comes down. This is repeated every 10 minutes until the internal anal sphincter is felt to close off again, or no stool has been passed after the last two stimulations. Alternatively, they can infuse a mini enema to stimulate the reflex. A few SCI maintain a deep nonspecific pelvic sensation of rectal fullness and can be taught to identify this sensation and then stimulate reflex emptying. Many patients can sense when defecation is complete.

Adequate hydration should be ensured and dietary and supplemental fiber should also be adequate. Hyperosmotic agents such as lactulose and sorbitol can contribute to facilitating bowel motility and regularity.

Sexual dysfunction following SCI. The person who suffers spinal cord injury is typically a fit young man at a sexually intense time of life. Most men following SCI are able to obtain reflex erections, but these are not usually adequate for intercourse and few experience psychogenic erections. The possibility of resuming sexual activity is a major concern of those injured and it has been shown that

the successfulness of rehabilitation is correlated with sexual potency. Although intracavernosal injections and vacuum pump devices were used by men with SCI, the introduction of Viagra, which increases erectile response within the context of sexual stimulation by their partner, has been greatly welcomed (see Chapter 4). A placebo-controlled trial showed sildenafil was highly effective in men with SCI in improving ability to achieve and maintain an erection and increase satisfaction with intercourse. It was effective even in those who did not have residual erectile function [22].

Very few men following SCI are able to ejaculate during intercourse, but if fertility is an issue, either a vibrator applied to the penis or electrical stimulation of the pelvic plexus by a rectal probe will induce ejaculation in most cases.

CONCLUSION

Patients with myelopathy frequently present with major problems with pelvic organ function. A comprehensive approach that integrates the assessment and treatment of bowel, bladder, and sexual dysfunction represents the strategy with the greatest likelihood of achieving satisfactory results. However, their caring physicians can institute effective diagnostic and treatment approaches that can enhance functional capability and ultimately promote a higher quality of life for patients with abnormalities of the genitourinary system. While the neurologist can successfully serve as the principal care provider for such patients, close collaboration with the urologist substantially adds value to the multidisciplinary management of those with spinal cord disease.

KEYPOINTS:

- Patients with myelopathy frequently present with major problems with pelvic organ function. A comprehensive approach that integrates the assessment and treatment of bowel, bladder, and sexual dysfunction represents the strategy with the greatest likelihood of achieving satisfactory results.

CASE-ORIENTED MULTIPLE CHOICE QUESTIONS

► A woman of 45 with secondary progressive MS complains of worsening bladder control. Despite taking 2.5 mg oxybutynin t.d.s. she had daily episodes of urge incontinence and wets the bed at night at least twice a week. The most appropriate next step in establishing more effective management is:

- A. Spinal cord MRI.
- B. Cystometry.
- C. Measurement of post micturition residual urine volume.
- D. Cystoscopy.
- E. Measurement of pudendal evoked potential.

The answer is C. Post micturition residual urine is a significant cause of symptoms of storage dysfunction. Oxybutynin will reduce the amplitude and frequency of unstable contractions and also increase bladder capacity; however, a persistent post micturition residual will decrease the time taken to fill the bladder to capacity.

► A man of 35 with a diagnosis of relapsing and remitting MS was recovering from a relapse in which there had been spinal cord demyelination with spastic paraparesis and lower limb weakness such that his mobility was reduced to 10 yards (9.1 meters) and he needed a stick to walk. Following a course of steroids, his walking improved somewhat, but he then presented with erectile dysfunction. Although morning erections were preserved, the patient was unable to obtain an erection sufficient for intercourse. The most appropriate initial step in management for the above case is:

- A. Investigation by a glucose tolerance test.
- B. Referral for psychosexual counseling.
- C. Consultation during which it is explained to him his ED is an expected result of spinal cord demyelination.
- D. MRI of the spinal cord.
- E. Nocturnal penile tumescence studies.

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The answer is C. When a patient with an established neurological diagnosis presents with ED there is no requirement to perform investigations, as the likely cause of the ED is neurological.

- A 40-year-old man with secondary progressive MS describes difficulty with urinary urgency, urge-incontinence and nocturia. There has been no recent history of urinary tract infections. Which of the following is the most important initial evaluation:

- A. Urinalysis.
- B. Renal ultrasound.
- C. Cystoscopy.
- D. Prostate specific antigen.
- E. Post-void residual urine measurement.

The answer is E. Again, post micturition residual measurement is the most important test to perform as the neurological diagnosis is certain, a residual is expected in this condition and would account for the symptoms.

- A 31-year-old woman presents with an 8-year history of multiple sclerosis. Her clinical course has been characterized by episodes of optic neuritis, transverse myelitis, and bilateral internuclear ophthalmoplegia. Over the last 6 months she has developed moderately severe lower extremity spasticity in association with involuntary jerks and spasms. Examination shows a spastic paraparesis, diffuse hyperreflexia, bilateral extensor plantar responses, bilateral ankle clonus, and a spastic gait. She complains of bowel urgency and also inability of evacuation. Which of the following accurately describes the underlying mechanism for her complaint:

- A. Constipation from poor neuro-gastrointestinal signaling.
- B. Immobility.
- C. Weak abdominal muscles.
- D. Reduced gastrocolic reflex.
- E. Pelvic floor muscular spasticity (recto-sphincter dyssynergia).

The answer is E. Patients with MS present with a wide variety of bowel complaints from constipation to fecal incontinence, especially of loose stools. However, in this case, the combination of fecal urgency and difficulty in evacuation indicate that pelvic floor muscular spasticity is the underlying problem.

- A 51-year-old woman with secondary progressive MS has recently ceased to have menstrual periods and now complains of hot flushes, depression, sexual dysfunction and fatigue. Her MS history includes two episodes of partial sensory transverse myelitis that left her with residual dysesthesias in the lower extremities and in the perineum. In terms of her sexual dysfunction, with a specific focus on treatment intervention, all of the following are important to explore initially except:

- A. Vaginal lubrication.
- B. Serum testosterone levels.
- C. Ability to become aroused and achieve orgasm (sensory threshold).
- D. Current use of antidepressants.
- E. Pain during sexual activity.

The answer is B. A raised testosterone level would be the most unexpected cause of female sexual dysfunction in this case, given that there is an established neurological diagnosis that would account for her symptoms.

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