

CHAPTER 1

A NEUROLOGIST'S CLINICAL AND INVESTIGATIVE APPROACH TO PATIENTS WITH BLADDER, BOWEL AND SEXUAL DYSFUNCTION

INTRODUCTION

Neurologists encounter many patients with symptoms that may be due to disorders of sacral innervation. These are either patients with established neurological disease who commonly have complaints of urogenital dysfunction or patients of whom the question is being asked “is there a neurological basis for the patient’s complaints of bladder, sexual and sometimes bowel dysfunction?” The approach to each group is necessarily different. For those patients known to have neurological disease, it is important that the neurologist be aware of the likely problems and be familiar with management and treatment options. In those in whom there is a question as to whether or not the complaint is part of a general neurological condition, the neurologist will need to know what investigations might be contributory.

Later sections of this teaching course deal in detail with causes and investigation of specific complaints, i.e. urinary incontinence and retention, constipation and fecal incontinence and sexual dysfunction. However, at this stage, a broad understanding of the neurological control of pelvic organs and how this may be affected by disease is needed.

HISTORY

Table 1 shows what symptoms might be expected from neurological disease at different levels. Clustering of symptoms is important in trying to decide if pelvic organ complaints are due to “ordinary”, local pathology or are neurogenic. For example, in a patient with spinal cord disease, bladder and sexual dysfunction are usually present together, whereas if bladder symptoms are due to prostatic outflow obstruction, sexual function is

preserved. Therefore, when taking the history, attention should focus on identifying whether a pelvic organ complaint is isolated or is part of a symptom complex.

CLINICAL NEUROLOGICAL EXAMINATION

The value of the clinical neurological examination is in recognizing co-existent general neurological problems. It is less useful in detecting defects of sacral innervation. This is where clinical neurophysiological investigations might have been expected to provide important information, but in practice, with one or two notable exceptions (see Pullout 1), is disappointingly not the case.

If a focussed clinical examination fails to detect evidence of neurological disease, laboratory investigations rarely provide a diagnosis.

The following sections describe briefly what the typical history and clinical examination might reveal in each condition.

Peripheral neuropathy. The neurologist may be asked if a patient’s peripheral neuropathy is the cause of their bladder dysfunction. Many neuropathies are length dependent, the maximum deficit being evident in the longest fibers, whereas the nerve fibers to the bladder are relatively short. If the innervation of the bladder has been affected as part of a generalized neuropathy, there is usually evidence of extensive disease with a long history of neuropathic symptoms, including sensory changes in the feet and sometimes the hands, with loss of pain and temperature perception with or without positive symptoms, such as painful dysesthesia. On examination, both knee and ankle jerks will be absent and small fiber sensory impairment demonstrable to the level of the ankles.

KEYPOINTS:

- The approach to each group is necessarily different. For those patients known to have neurological disease, it is important that the neurologist be aware of the likely problems and be familiar with management and treatment options. In those in whom there is a question as to whether or not the complaint is part of a general neurological condition, the neurologist will need to know what investigations might be contributory.
- Clustering of symptoms is important in trying to decide if pelvic organ complaints are due to “ordinary”, local pathology or are neurogenic.
- The value of the clinical neurological examination is in recognizing co-existent general neurological problems. It is less useful in detecting defects of sacral innervation.
- If the innervation of the bladder has been affected as part of a generalized neuropathy, there is usually evidence of extensive disease with a long history of neuropathic symptoms, including sensory changes in the feet and sometimes the hands, with loss of pain and temperature perception with or without positive symptoms, such as painful dysesthesia.

NEUROLOGIC BLADDER, BOWEL AND SEXUAL DYSFUNCTION

KEYPOINTS:

- It is exceptional for there to be single pelvic organ dysfunction due to a cauda equina lesion and corroborative evidence of other sacral root symptoms should be sought before, for example, attributing isolated bladder symptoms to a partial lesion.

TABLE 1 Conjunction of pelvic organ symptoms which may occur with neurological disease at different levels in the nervous system

Neurological lesion	Symptoms of pelvic organ dysfunction
Innervation within the pelvis	Bladder emptying difficulties ED, sometimes FSD
Peripheral neuropathy	ED (early) Bladder emptying difficulties (late) Diarrhea Postural hypotension
Cauda equina	Saddle sensory impairment Stress urinary incontinence Difficulty in initiation of micturition Urgency (occasionally) Sexual sensory loss ED, FSD Constipation Fecal incontinence/difficulty in evacuation
Spinal	Somatic sensory level Urinary urgency Incomplete bladder emptying ED, FSD Difficulty in bowel evacuation (in advanced disease)
Pontine (very rare)	Internuclear ophthalmoplegia Urinary retention
Extrapyramidal	Parkinsonism (advanced in IPD, minor in MSA) ED (early in MSA) Urinary incontinence (early in MSA) Constipation
Frontal	Personality change Urinary urge incontinence Fecal incontinence (exceptional)

ED, erectile dysfunction; FSD, female sexual dysfunction; IPD, idiopathic Parkinson's disease, MSA, multiple system atrophy.

Bladder or bowel symptoms should not be attributed to peripheral neuropathy unless there are other features of autonomic involvement in diabetics. However, sexual dysfunction, particularly erectile failure, occurs early in diabetes and may in fact be a presenting symptom when there is little or no evidence of generalized neuropathy to find on examination.

Cauda equina. Cauda equina lesions are likely to cause bladder and sexual complaints as well as difficulties of defecation. It is exceptional for there to be single pelvic organ dysfunction due to a cauda equina lesion and corroborative evidence of other sacral root symptoms should be sought before, for example,

attributing isolated bladder symptoms to a partial lesion.

Cauda equina disease or damage that affects only S2-S4 will produce sensory loss restricted to the perineum and the back of the thighs. A lax anal sphincter may be demonstrable and sacral reflexes, such as the bulbocavernosus reflex, lost. However, even with extreme caudal spinal lesions, there are usually also neurological abnormalities in the lower limbs, and foot deformities due to weakness of the intrinsic muscles may be present if the problem is long standing.

Examination of the lower back and sacrum is very important in these cases.

PULLOUT 1 Clinical neurophysiological investigations of the pelvic floor

Bulbocavernosus reflex

A neurophysiological method for recording the bulbocavernosus reflex, which had been clinically regarded to be of value in assessing patients with neurogenic bladder disorders, was first reported in 1967. Although abnormal in a proportion of patients with neurological disease causing pelvic organ dysfunction, the test was of little value when applied to patients with uncertain neurological lesions presenting with hypocontractile bladders or ED. The responses are mediated by large myelinated fibers and the small myelinated or unmyelinated fibers that either innervate the smooth muscle or constitute the functionally important afferent nerve supply of the region, are not tested.

The bulbocavernosus reflex was used extensively for the diagnosis of neurogenic erectile dysfunction. Elicited clinically by squeezing the glans penis and neurophysiologically by an electrical pulse to the dorsal penile nerve, a contraction of the bulbocavernosus muscle was observed or recorded electromyographically. The reflex tested the integrity of large myelinated nerve fibers in the S2-S4 segments. The minimum latency response was measured and values greater than 45 ms were considered abnormal. Although many patients with neurogenic ED due to either cauda equina or lower motor neuron lesions had abnormally prolonged or absent BCR responses, the test was not sensitive and some men with established cauda equina lesions or diabetic peripheral neuropathy had normal responses. Furthermore, the response was prolonged in some men with a demyelinating neuropathy who had unimpaired sexual function.

Terminal motor latency of the pudendal and perineal nerves

Measurement of the terminal motor latency of the pudendal and perineal nerves (PTML) was a technique devised at St Mark's Hospital, London. It has been used to demonstrate pathophysiological changes in these nerves in women with fecal, urinary or double incontinence.

The pudendal nerve is stimulated transrectally near the ischial spine through the wall of the rectum or vagina using an electrode mounted on the tip of the examiner's finger. An electrode is mounted at the base of the finger which records from the anal sphincter and a ring electrode mounted on a Foley catheter can be used to record from the periurethral striated muscle. The latency of the response has been found to be prolonged in women with urinary stress incontinence following childbirth and women with fecal incontinence due to sphincter weakness. It is thought that stretching of the nerves during parturition and also with straining at defecation in chronic constipation results in pudendal nerve injury. Although of considerable research value, this test is not used in the routine assessment of women with urinary stress incontinence, nor has it proved to be as useful as electromyography of the anal sphincter in the assessment of fecal incontinence.

Pudendal evoked response

The pudendal evoked potential (PEP) is easily recorded using a similar technique to that which is used for recording the tibial evoked potential, but with stimuli applied to the dorsal nerve of the penis or clitoris. The response has a similar waveform and even latency to that of the tibial evoked response — the slower conduction being due to the composition of the penile nerve being sensory only and lacking the fast conducting muscle afferent fibers of the tibial nerve.

At one time, it was recommended that the pudendal evoked response be included as part of screening tests in the investigation of erectile dysfunction. The PEP is abnormal in patients with spinal cord disease including MS, but is less sensitive in detecting spinal cord disease than is a comparison of the tibial evoked potentials recorded from both limbs. Furthermore, spinal cord dysfunction is usually evident from clinical examination.

Pullout 1 continued ►

Pullout 1 continued ►

Anal or urethral sphincter EMG

Recording from the striated muscle of the urethral sphincter or anal sphincter during cystometry was used as a means of detecting inappropriate sphincter contraction during detrusor contraction, the disorder known as detrusor sphincter dyssynergia. However, for several reasons, this type of kinesiological EMG recording is now little used, although sphincter EMG performed as a separate neurophysiological test remains a valuable investigation in some circumstances.

Needle electrode EMG of either the anal or urethral sphincter can be performed to show evidence of sacral segment or root damage in much the same way as EMG is used at somatic sites. However, because motor units in the sphincter fire tonically, it is difficult to recognize changes of denervation and most often changes of reinnervation are sought, based on the analysis of individual motor units captured using a trigger and delay line. In general, EMG is held to be the most valuable of the pelvic floor investigations to detect lower motor neuron damage. EMG of the striated musculature of the pelvic floor can demonstrate changes of denervation and chronic reinnervation in patients with cauda equina lesions as well those with suspected MSA. The striated muscles of the sphincters are innervated by anterior horn cells which lie in Onuf's nucleus in the sacral part of the spinal cord and neuropathological studies showed loss of cells in Onuf's nucleus in patients dying with Shy-Drager syndrome. These changes may be reflected by abnormalities of sphincter EMG in life. Changes of reinnervation in MSA are non-specific and some caution must be exercised in interpreting EMG findings in multiparous women or patients who have had extensive pelvic surgery. There is some debate as to the value of the test in distinguishing between MSA and IPD, but extreme prolongation of the mean of 10 motor units in a patient with minor parkinsonism and severe urinary incontinence and ED is strongly indicative of MSA. The other condition in which urethral sphincter EMG has proved to be of particular value is the investigation of young women in urinary retention.

Unfortunately, there is as yet no neurophysiological means of investigating detrusor smooth muscle function.

Autonomic function testing

Using a combination of cardiovascular tests, generalized sympathetic and parasympathetic failure can be recognized, which may be associated with urogenital dysfunction. Cardiovascular autonomic function tests measure changes in heart rate variability and blood pressure in response to deep breathing, changes in posture from lying to standing, cold stimuli, isometric exercise, sudden inspiratory gasps or Valsalva maneuver. These tests investigate only generalized cardiovascular autonomic failure and overlook possible focal or regional abnormalities in peripheral or central autonomic nerve function. It is not surprising, therefore, that normal cardiovascular results are found in some patients with abnormal erectile function and vice versa.

Sympathetic skin responses

The changes in skin resistance that occur following various internally generated or externally applied arousal stimuli result from an increase in sweat gland activity mediated by the sympathetic nervous system, and lead to a change in skin voltage that can be recorded using two surface electrodes. This response is called the sympathetic skin response (SSR) and is thought to originate from synchronized activation of sweat glands in response to a discharge of the efferent sympathetic nerve fibers. SSR responses recorded from the limbs can be used in the detection of mixed axonal neuropathies and have also been measured on the genital skin. The SSR could be recorded from the genitalia in normal subjects, but the response was absent in some diabetics with erectile dysfunction, both with and without a previously diagnosed neuropathy.

“Corpus cavernosum EMG”

The electrical activity that can be recorded from the corpus cavernosum using either a concentric needle electrode or surface electrodes has been called “corpus cavernosum electromyography” (CC-EMG). Activity is recorded on a compressed time base with the subject at rest. Studies on “CC-EMG” have recognized that the recorded potentials are sympathetically mediated and that mental stimulation increased the frequency of the potentials. The similarity between “CC-EMG” and activity that can be recorded from surface electrodes placed on the limb of a patient using the set up as used to record SSR, but on a slow time base, suggests the two types of activity may have a common genesis, although there are some differences. In general, there is good evidence that these recordings represent some form of autonomic activity from the genitalia.

Spinal cord. Because of the relative levels of innervation of the lower limbs and the pelvic organs, it is unusual to have a lesion between the pons and the sacral part of the cord giving rise to a neurogenic bladder or sexual dysfunction that does not also produce signs of an upper motor neuron lesion in the lower limbs. This is undoubtedly the case in patients with multiple sclerosis (MS) but the rule appears to hold for most other instances of spinal pathology, unless the lesion is central, intramedullary and small.

The majority of men with spinal cord pathology causing bladder symptoms have impaired erectile function, but bowel complaints would not be an expected part of the picture, unless there is a marked neurological deficit. Symptoms and signs of a paraparesis of variable severity should also be expected.

Brainstem. Brainstem or pontine pathology usually causes marked neurological deficits, but occasionally a lesion can be sufficiently dorsal and discreet to produce predominantly a defect of bladder function. An internuclear ophthalmoplegia may be an accompanying sign in such cases, due to the proximity of the median longitudinal fasciculus.

Extrapyramidal. Parkinsonian features are marked in patients with long-standing Parkinson’s disease who develop genitourinary symptoms as part of the neurological picture. Neurological abnormalities in patients with multiple system atrophy (MSA), however, may be subtle at a time when genitourinary symptoms present and it is important to examine for early cerebellar signs and also postural hypotension.

Supra pontine. The contribution of supra pontine pathology to neurogenic bladder dys-

function, with the exception of areas in the frontal lobes is poorly defined. Patients with frontal lobe incontinence may have neuropsychological impairment, such as a change of personality, but are not indifferent to their incontinence unless there has been extensive frontal lobe damage.

NEUROPHYSIOLOGICAL INVESTIGATIONS OF THE PELVIC FLOOR

Various neurophysiological investigations of the pelvic floor and the sphincters have been developed and used over the years, but the current view is that few are of diagnostic value. Although there have been many research papers reporting abnormalities of neurophysiological investigations in patients with established neurological disease, studies of the sensitivity of these compared with the neurological examination were rarely performed. Exceptions are the use, but disputed value of, anal sphincter EMG to recognize changes of chronic reinnervation in multiple system atrophy and the use of urethral sphincter EMG to recognize a primary disorder of sphincter relaxation in young women with isolated urinary retention.

Because, historically, neurophysiological testing has been thought to be important in the past, a brief description is given here (Pullout 1). Greater detail can be found in a review articles [1, 2].

IMAGING OF THE NERVOUS SYSTEM

Neurological imaging may be important if the patient had been referred from a urologist to a neurologist asking if the patient has a neurogenic basis for their symptoms. Imaging is

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- Various neurophysiological investigations of the pelvic floor and the sphincters have been developed and used over the years, but the current view is that few are of diagnostic value.

NEUROLOGIC BLADDER, BOWEL AND SEXUAL DYSFUNCTION

KEYPOINTS:

- In a patient with bladder symptoms and established neurological disease, urodynamic investigations may be performed to understand the pathophysiological basis for the patient's complaint and obtain information on which to base recommendations for management of incontinence.
- A change in bowel habit unrelated to major changes in neurological condition or rectal bleeding always warrants investigation to exclude bowel malignancy.
- Now that there are simple and effective symptomatic treatments for erectile dysfunction (ED), the emphasis has moved away from investigations that were carried out in former times to try and distinguish between organic and psychogenic causes of ED. In men with established neurological disease known to cause ED, laboratory investigation is not indicated, unless the response to treatment is poor.

particularly indicated to exclude a supra pontine abnormality or a sub-sacral lesion, and magnetic resonance imaging is now the investigation of choice.

Neurological imaging has no role in determining management of pelvic organ dysfunction in a patient with a known neurological diagnosis.

INVESTIGATIONS OF BLADDER SYMPTOMS

Urodynamics. In a patient with bladder symptoms and established neurological disease, urodynamic investigations may be performed to understand the pathophysiological basis for the patient's complaint and obtain information on which to base recommendations for management of incontinence. The term "urodynamics" encompasses any investigation of urinary tract function, although it is often used incorrectly as a synonym for cystometry.

The role of urodynamics in trying to decide if a patient has a neurogenic bladder disorder is limited. The majority of patients sent by urologists to neurologists have been demonstrated on filling cystometry to have bladder overactivity (see Chapter 2), and because there are non-neurogenic causes for this, the neurologist must try to confirm or refute that there is neurological basis for the problem. The clinical neurological examination is critical for this.

Ultrasound scanning of the urinary tract. Ultrasound scanning has largely overtaken intravenous urography as the method of choice for examining the upper renal tract to detect dilatation, although urologists still prefer intravenous urography to look for ureteric stones.

The residual urine volume left after voiding is a critical factor in determining manage-

ment of neurogenic incontinence and can be measured by non-urologists using a small relatively inexpensive ultrasound scanner (see Chapter 2).

INVESTIGATION OF CONSTIPATION/FECAL INCONTINENCE

Bowel complaints do occur in patients with neurological disease which can mostly be managed symptomatically. However, the contribution of neurological disorders to bowel control is not an area that is well understood and investigations may be required in individual patients to define the pathophysiology of their symptoms as well as exclude non-neurological causes of fecal incontinence or co-incidental disease. A change in bowel habit unrelated to major changes in neurological condition or rectal bleeding always warrants investigation to exclude bowel malignancy. A description is given in Chapter 3 of the various investigations that can be performed to study function and structure.

INVESTIGATION OF SEXUAL DYSFUNCTION

Now that there are simple and effective symptomatic treatments for ED, the emphasis has moved away from investigations that were carried out in former times to try and distinguish between organic and psychogenic causes. In men with established neurological disease known to cause ED (see Table 1), laboratory investigation is not indicated, unless the response to treatment is poor.

In the general population, ED often has a significant vascular component. This type of problem may be investigated using a variety of techniques to study penile hemodynamics, as described in Chapter 4 (Pullout 2).

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