CHAPTER 7

PERIPHERAL NEUROPATHIES

KEYPOINTS:

- Which of these types of peripheral neuropathies produce bladder, bowel or sexual dysfunction? The general rule is that those polyneuropathies that include concomitant involvement of the autonomic nerves, and focal neuropathies of the nerves of the pelvis, are the ones that cause such symptoms.
- Diabetes is the commonest cause of polyneuropathy in most countries.

INTRODUCTION

Peripheral neuropathy is a very broad term. Its definition is based on what constitutes the peripheral nervous system, and what types of damage and dysfunction can occur to these nerves.

The peripheral nervous system is divided into the *somatic* and the *autonomic* nervous systems (ANS). *Somatic* motor nerve fibers control skeletal muscle function, while the sensory fibers mediate cutaneous and some deep sensations. The *autonomic* sensory and motor nerve fibers control most aspects of cardiovascular, gastrointestinal, bladder, and sexual functions.

Disorders of peripheral nerves (*peripheral neuropathies*) can be classified in many ways. One fundamental consideration is whether the neuropathy involves the peripheral somatic nerves exclusively, the autonomic nerves only, or a combination of the two. Another fundamental classification is into clinicopathological patterns of nerve damage: polyneuropathy (generalized peripheral neuropathy), focal peripheral neuropathy, and mononeuropathy multiplex (Table 1).

In a *polyneuropathy*, the peripheral nerves are affected symmetrically, and usually the longest nerve fibers are damaged first and maximally. Thus the symptoms and signs involve both feet first, and as the disorder progresses, the hands are also both involved.

A *focal peripheral neuropathy* is a discrete lesion to a nerve, be it a spinal nerve root, a plexus, an individual major nerve trunk, or a branch from such a nerve. The neurological deficit is restricted to the motor and sensory territories supplied by the damaged nerve.

Mononeuropathy multiplex refers to the situation in which two or more individual nerves or branches are involved. Thus the symptoms and signs are restricted to the territories of these damaged nerves.

Which of these types of peripheral neuropathies produce bladder, bowel or sexual dysfunction? The general rule is that those polyneuropathies that include concomitant involvement of the autonomic nerves, and focal neuropathies of the nerves of the pelvis, are the ones that cause such symptoms. Thus the neurologist who thinks that the patient's sphincteric dysfunction is caused by a peripheral neuropathy, has to first ask the question: is this a polyneuropathy or focal pelvic nerve damage? The next question is: if this is a polyneuropathy, what is the cause? There are many causes of polyneuropathy, but there are relatively few that cause prominent bowel, bladder, or sexual dysfunction (Table 2). In terms of focal pelvic nerve damage, the cause may be obvious or may require investigations to show the site and type of damage.

TABLE 1 **The major types of peripheral neuropathies**

Polyneuropathy (generalized peripheral neuropathy)
Focal peripheral neuropathy
Mononeuropathy multiplex

TABLE 2 **Polyneuropathies causing bladder, bowel and sexual dysfunction**

Diabetes mellitus
Amyloidosis
Porphyria
Guillain-Barré syndrome
Other chronic neuropathies

DIABETES MELLITUS

Diabetes is the commonest cause of polyneuropathy in most countries. It is also by far the commonest polyneuropathy to be associated with bowel, bladder, and erectile dysfunction. Diabetes mellitus gives rise to a variety of different types of peripheral neuropathies, but the most frequent is a polyneuropathy. The severity and manifestations of this are highly variable from being completely unnoticed

CASE 1

History: This 35-year-old man has had type I diabetes mellitus since age 13 (for 22 years). He has had retinopathy for 5 years, treated with laser techniques, and nephropathy for 4 years. Polyneuropathy was recognized at age 30, and he developed a Charcot joint of the left ankle at age 34.

Autonomic dysfunction began at age 25 with erectile dysfunction (ED) which was managed with an implant. He then developed intermittent diarrhea. He was occasionally constipated. Bladder function has remained normal. Orthostatic hypotension on standing up from bed has been troublesome, so he sits before standing. Syncopal episodes have occurred in a variety of situations and for no obvious reason, and with no consistent warning symptoms. Sweating is also abnormal: he sweats heavily from the neck down to the mid-thighs, but is dry everywhere else.

Examination: There was moderate distal wasting and weakness in the limbs, and complete areflexia. Light touch and pin prick were diminished in the tips of the fingers and in the feet to the mid-calf. Vibration was absent in the toes, but present at the ankles; proprioception was absent in the toes. Blood pressure was 160/90 mm Hg lying and 100/60 mm Hg standing. His pulse was 90 per minute and regular in both positions. His hands and feet were completely dry, but when sitting in a warm examination room, his trunk was excessively sweaty and his shirt became completely wet.

Management: The ED was managed with an implant, diarrhea with diphenoxylate hydrochloride, and the orthostatic hypotension with fludrocortisone.

Comment: This young type I diabetic had moderate to severe polyneuropathy and had sustained a Charcot joint in one foot. He also had DAN manifested by chronic diarrhea, orthostatic hypotension, fixed tachycardia (lack of vagal innervation to the heart), and sweating abnormalities. As is often the case, the ED developed earlier than other autonomic abnormalities. The lack of any symptoms of bladder dysfunction shows how patchy the autonomic damage in diabetes can be. The appearance of these features of DAN are very worrying because of the poor prognosis associated with this condition.

by the patient to a severe disabling neuropathy with marked wasting and weakness and impaired gait and hand function. Fortunately, severe neuropathies are relatively uncommon.

Diabetic autonomic neuropathy (DAN) is a frequent accompaniment of severe diabetic polyneuropathy (see Cases 1 and 3) [1]. However, sometimes DAN is the dominant peripheral nerve involvement. The usual diabetic to run into bladder, bowel, and sexual dysfunction secondary to autonomic neuropathy has had longstanding type I or II diabetes, and also has retinopathy and some degree of nephropathy (see cases). It is important to note that DAN can be restricted to certain organs or functions, or may be widespread, involving most or all of the peripheral ANS.

Bladder dysfunction. Bladder dysfunction in diabetics ("diabetic cystopathy") has been long recognized [2]. The precise incidence is uncertain because there are no community-based surveys; in selected series, the incidence ranges from 5 to 71%. The onset is usually insidious. Initially, there is reduced sensation of bladder fullness, and decreased frequency of voiding. This is followed by slowing of the urinary stream and difficulty in voiding, so that the patient voids by abdominal straining (see Case 2). Postvoiding dribbling may also occur. Bladder neck obstruction caused by benign prostatic hypertrophy produces many of the same symptoms. Urological studies including pressure-flow studies made during cystometry are required to distinguish between these entities.

The impaired bladder emptying and urinary retention predispose to urinary tract infections. These, in turn, lead to secondary bladder abnormalities, such as fibrosis and scarring. These further impair bladder function, leading to more urinary tract infections and the complications thereof.

Bowel dysfunction. Three types of bowel dysfunction occur in diabetics: constipation, diarrhea, and anorectal incontinence [3]. Of these, constipation is probably the commonest, and can be severe. Diabetic diarrhea can also be very troublesome. This, like erectile dysfunction, may be an isolated symptom of

restricted autonomic dysfunction, other abnormalities of autonomic function being absent. It typically occurs at night or following meals. It may be explosive and so fecal incontinence may be a feature. Diabetic diarrhea can be chronic, but is often intermittent and may alternate with bouts of constipation or with normal bowel movements. Patients may tread the fine line between constipation and diarrhea (see Case 3). Medications used to control one can precipitate the other. Diarrhea may be confused with true anorectal incontinence which is due to poor anal sphincteric function, not diarrhea per se. This is by far the most difficult of the three diabetic bowel symptoms to deal with.

Diabetics may have dysfunction of the more proximal gastrointestinal (GI) tract [4]. Esophageal motility studies are frequently abnormal, but symptoms are uncommon. When present, they consist of heartburn and dysphagia. Gastric atony may be an isolated radiographic finding or may cause abdominal distension (bloating), nausea, vomiting (Cases 2 and 3). These upper GI tract symptoms may occur separately or in addition to the bowel symptoms.

The GI dysfunction in diabetics is principally due to impaired function of the enteric nervous system that controls many aspects of GI physiology [4]. Thus, diabetics have abnormalities of bowel motility, impaired regional blood flow control, reduced neurally mediated afferent signals, impaired water and electrolyte absorption, reduced intestinal and pancreatic enzyme secretion, gallbladder dysfunction and abnormal bile acid secretion. An important additional factor is bacterial colonization of the normally sterile small intestine.

Sexual dysfunction. This is unfortunately common in diabetic males, many series showing a prevalence of 30-60% (see Case 1) [5]. Erectile dysfunction (ED) is much more frequent than ejaculatory difficulties. The latter usually involves retrograde ejaculation rather than total lack of ejaculation.

Erectile dysfunction in diabetic men usually begins insidiously with a progressive decline in erection rigidity and duration, to the point where penetration and intercourse become impossible. Libido is unaffected. If sexual stimulation is engaged in, despite the lack of erection, ejaculation and orgasm often occur, indicating the particular susceptibility

CASE 3

History: This 38-year-old woman has had type I diabetes mellitus for 15 years. Non-neurological diabetic complications have included retinopathy requiring laser treatments, osteomyelitis of a foot, and declining renal function.

For the past 4 years, she has had paresthesias then numbness in the feet, but no sensory symptoms in her hands or limb weakness. For 6 years, she has had chronic diarrhea, at times explosive, and sometimes leading to fecal incontinence. She was found to be lactose-intolerant, but treatment for this did not improve the diarrhea. Bladder function has been normal. Over the past 4 years, she has developed progressively worsening orthostatic hypotension. Even with treatment, this has markedly limited her ability to walk and do many simple activities of daily living. The most recent symptoms have been abdominal bloating, nausea and vomiting.

Examination: The upper limbs were neurologically normal. In the legs, there was mild weakness of distal muscles and areflexia. Light touch and pin prick were diminished to the mid-calves. Vibration was present, but for a short duration in the toes; proprioception was normal. Blood pressure lying was 110/70 mm Hg and standing 80/50 mm Hg, with no increase in heart rate.

Management: Many treatments, including antibiotics and medications to reduce bowel motility, were tried for her diarrhea. These were generally ineffective or led to constipation, so the diarrhea remains a persistent problem. Antiemetic medications, combined with small frequent meals, have helped to control the upper GI symptoms. The orthostatic hypotension has been very difficult to treat. Fludrocortisone and midodrine, combined with non-pharmacological maneuvers, such as elastic stockings and head-up tilt of her bed, have been of some help.

Comment: This diabetic has mild to moderate polyneuropathy, but some severe manifestations of DAN. As in Case 2, this patient illustrates the discrepancy that may occur between involvement of the somatic (in this case, minimal) and the autonomic (substantial) peripheral nervous systems in diabetic patients. Unfortunately, this patient's dysautonomic symptoms have been very difficult to control adequately.

KEYPOINTS:

- ED is frequently present in diabetic men who may have mild somatic polyneuropathy and no manifestations of dysautonomia except for ED.

of the erectile mechanisms in this disorder. Most of these patients report a cessation of spontaneous nocturnal erections.

Unlike the bladder and most types of GI dysfunction in diabetics (except diarrhea), ED is frequently present in diabetic men who may have mild somatic polyneuropathy and no manifestations of dysautonomia except for ED. It is now thought that the high prevalence of ED and its relatively poor response to sildenafil may be due to a metabolic effect which blocks the vasodilatator action of released nitric oxide in corporeal tissue. The contribution due to involvement of the autonomic nerves controlling erection is difficult to determine because specific tests of these nerves are lacking.

Sexual function in female diabetics has only been investigated to a limited extent [6]. Reduced vaginal secretion on sexual arousal has been reported.

The impact of DAN. The mortality rate in diabetics with DAN after 5 years has been reported to be as high as 56% [1]. Another 5-year survival study has shown that the mortality rate in patients with DAN is increased 5-fold compared with those without DAN [7]. Postural hypotension, gastroparesis, and hypoglycemic unawareness correlate well with abnormalities of cardiovascular reflexes and carry a poor prognosis. Diarrhea and erectile impotence are often not associated with abnormal tests of autonomic function and, when present as isolated symptoms, do not carry a poor prognosis.

It is not yet clear whether DAN is directly responsible for these extra deaths, whether it is one of several contributing factors, or is a marker of widespread diabetic complications. However, one study has shown that DAN *is* an independent risk factor for excessive cardiovascular mortality in type II diabetics [8]. Diabetics appears to have a higher incidence of sudden death compared to non-diabetics, with cardiac arrhythmias from DAN being a possible contributing cause [1].

AMYLOID NEUROPATHY

Amyloid is a proteinaceous substance that becomes deposited in various tissues in certain disease states or as the result of a gene abnormality. There are a variety of types of

amyloidosis, but those manifesting as somatic and autonomic neuropathy are: (a) immunoglobulin amyloidosis; and (b) certain types of familial amyloidosis.

Immunoglobulin amyloidosis (now called AL, after the light chain proteins that constitute the amyloid) occurs either as a primary disorder, or associated with multiple myeloma, Waldenstrom's macroglobulinemia, or non-Hodgkin's lymphoma [9]. The dysautonomia often accompanies, but may precede, the manifestations of somatic polyneuropathy. GI motility abnormalities can produce pseudo-obstruction, troublesome diarrhea, or constipation, or each of these alternating with the other. Bladder dysfunction is common, as is ED in males. The other features of dysautonomia, such as orthostatic hypotension, anhidrosis, and pupillary abnormalities, also occur in these patients. The somatic polyneuropathy is initially of the *"small fiber type"*: Painful paresthesiasis are, therefore, prominent, and the examination shows a peripheral loss predominantly of pain and temperature sensations. There is later involvement of motor function and of the sensory modalities subserved by large myelinated fibers. Carpal tunnel syndrome occurs in about a quarter of the patients. The clinical course is relentlessly progressive with death within 5 years of the diagnosis due to renal, cardiac, or gastrointestinal involvement by amyloid. The diagnosis is made by finding amyloid deposits in biopsies of nerve, rectal mucosa, or abdominal wall adipose tissue.

A wide variety of dominant *hereditary/familial amyloidoses* have been described [10]. Those that involve peripheral nerves are called *familial amyloidotic polyneuropathy* (FAP) and are further classified on the basis of the specific mutation in the transthyretin gene. Over forty disease-causing mutations have been identified, with autonomic involvement occurring in about half. As in AL, the features are those of progressive polyneuropathy, dysautonomia, and, in some types, carpal tunnel syndrome.

PORPHYRIA

The porphyrias are hereditary disorders affecting hepatic heme metabolism. The four types with neurological manifestations are acute intermittent porphyria, variegate porphyria, hereditary coproporphyria, and δ-aminolevulinic acid dehydratase deficiency [11]. Patients can have acute attacks of predominantly motor and often proximal neuropathy, in conjunction with psychiatric features and abdominal pain; these latter features often precede the neuropathy. Constipation, intestinal stasis and dilatation, micturition difficulties, orthostatic hypotension, paroxysmal hypertension and tachycardia may all be part of the clinical syndrome. This disorder and these clinical manifestations are fortunately rare.

GUILLAIN-BARRÉ SYNDROME

Autonomic dysfunction occurs in about 80% of patients with Guillain-Barré syndrome (GBS) [12]. This takes the form of hyperactivity or hypoactivity of sympathetic and parasympathetic functions.

Major fluctuations in blood pressure are common, consisting of both hypertension and hypotension.

Sustained sinus tachycardia is common. Less frequent is bradycardia, but this is more serious since it sometimes leads to sinus arrest. A variety of other cardiac arrhythmias can occur, and may be responsible for sudden death in some patients. Anhidrosis occurs in the extremities; facial flushing is another uncommon manifestation of dysautonomia.

Bladder dysfunction occurs in approximately 20% of patients, though this is difficult to establish with certainty, because these patients are often catheterized for ease of micturition management rather than for bladder dysfunction per se. Cystometric studies show a delay in the initial sensation of bladder filling, and detrusor areflexia. Recovery of bladder dysfunction sometimes takes months. Constipation is common, and paralytic ileus occasionally occurs. Dysautonomic abnormalities are at their maximum during the peak period of paralysis. There does not seem to be any particular prognostic significance to these dysautonomic abnormalities in terms of the ultimate outcome of the patients.

MISCELLANEOUS CHRONIC NEUROPATHIES

Bladder, bowel and sexual dysfunction are fortunately rare in most hereditary neuropathies, such as Charcot-Marie-Tooth neuropathy (hereditary sensorimotor neuropathy),

KEYPOINTS:

- Autonomic dysfunction occurs in about 80% of patients with Guillain-Barré syndrome.

KEYPOINTS:

- Bladder and erectile dysfunction following treatment for prostate disorders - benign prostatic hypertrophy and prostatic cancer - merit special attention because of the frequency of these disorders, and indeed of these complications.
- "Nerve-sparing" surgical techniques involve identifying and preserving the nerves to the corpora cavernosa. These travel outside and behind the prostatic capsule in the lateral pelvic fascia until nearer the apex of the prostate when they lie just lateral to the urethra. It is at this point that they are particularly vulnerable to surgery. They then pass behind the penile artery and dorsal penile nerve to enter the corpora on each side. Bilateral nerve sparing leads to less ED than unilateral nerve sparing, which causes less ED than no nerve sparing

chronic inflammatory demyelinating neuropathy (CIDP), and neuropathy associated with monoclonal antibodies. The same holds true for patients with chronic axonal neuropathies, in many of which there is no definable cause.

FOCAL PELVIC NEUROPATHIES

Bladder, bowel and sexual dysfunction can occur from damage to the nerves innervating the pelvic organs, anywhere in the course of these nerves through the cauda equina, the spinal nerve roots, the sacral plexus, or to the various individual nerves that arise from the plexus. Cauda equina lesions are discussed in Chapter 6.

Fractures of the pelvic girdle, gunshot or other missile injuries, can cause damage to these nerves at any or at combinations of these sites. However, most injuries to these nerves are iatrogenic (Table 3).

Extensive pelvic surgery, such as abdominoperineal resection for rectal cancer, radical hysterectomy, and aortoiliac surgery are all likely to damage the pelvic parasympathetic nerves to the bladder and genitalia. A variety of types of voiding dysfunction and erectile dysfunction can result. Anorectal incontinence is fortunately rare.

Bladder and erectile dysfunction following treatment for prostate disorders — benign prostatic hypertrophy and prostatic cancer — merit special attention because of the frequency of these disorders, and indeed of these complications. Following transurethral prostatectomy (TURP) for benign prostatic hypertrophy (BPH), urinary retention or incontinence may occur immediately postoperatively, but improves markedly within weeks, and the long-term incidence is about 1% [13,14]. Some reports claim that ED occurs in up to 10% of men following TURP [15], but a prospective study in which erectile function was assessed pre- and postoperatively with nocturnal tumescence studies, showed that ED occurred in none of the 40 men studied [16]. Retrograde ejaculation is very common following TURP, but is usually accepted as a minor problem.

TABLE 3 **Surgical procedures producing injuries to pelvic nerves**

Abdominoperitoneal resection
Radical hysterectomy
Aortoiliac surgery
Prostatectomy
• Transurethral
• Radical

The situation is quite different for patients undergoing treatment for prostate cancer. The two standard treatments for this are radiotherapy or radical prostatectomy; the latter involves removal of the prostatic capsule to which the corporeal nerves are intimately bound. A meta-analysis of ED following these treatments has shown that chances of developing this complication after these two types of treatments were 30 and 60%, respectively — very important differences [17]. "Nerve-sparing" surgical techniques involve identifying and preserving the nerves to the corpora cavernosa. These travel outside and behind the prostatic capsule in the lateral pelvic fascia until nearer the apex of the prostate when they lie just lateral to the urethra. It is at this point that they are particularly vulnerable to surgery. They then pass behind the penile artery and dorsal penile nerve to enter the corpora on each side. Bilateral nerve sparing leads to less ED than unilateral nerve sparing, which causes less ED than no nerve sparing [18]. This post-prostatectomy ED is due to nerve, arterial or venous damage, or combinations thereof [19].

Micturition abnormalities are common following radical prostatectomy, the chief problem being incontinence of one sort or another. Estimates of the incidence vary between 2 and 87% [20]. Urodynamic studies have shown that in most affected patients, this is due to direct damage to the intrinsic urethral sphincter; a small minority have detrusor dysfunction due to nerve injury [20].

Based on finding prolongation of measurement of the terminal pudendal motor latency, it was thought that denervation of the sphincters and pelvic floor was an important cause of stress urinary incontinence and idiopathic fecal incontinence (see Chapter 1). It was suggested that the pudendal nerve was damaged during vaginal delivery and the problem further exacerbated by chronic straining at stool which caused pelvic descent and traction injury to the nerves. Since then, ultrasound studies have demonstrated that damage to the smooth muscle of the internal anal sphincter is a common finding in postpartum fecal incontinence [21], and stress urinary incontinence is thought to be due to direct damage to the striated muscle and fibroelastic supporting tissues of the pelvic floor.

DIAGNOSTIC APPROACHES

Clinical assessment. The history and examination are often sufficient for the neurologist to make a diagnosis of peripheral nerve disease as the cause of bladder, bowel, or erectile dysfunction. A key factor in the few polyneuropathies that are associated with these symptoms is that the patients often have motor and sensory symptoms in the hands and feet. In those who do not have such symptoms, a careful examination of distal strength, reflexes and sensation will reveal abnormalities characteristic of a polyneuropathy.

The clinical evaluation of the autonomic nervous system (ANS) is often not a usual part of a neurologist's appraisal, but is indicated here. Symptoms of orthostatic hypotension, upper GI tract dysfunction, such as abdominal bloating, dry eyes, dry mouth, sweating abnormalities, all additional indicators of disordered autonomic function. The examination of the ANS is restricted, but, nonetheless, often very revealing. Simply detecting a resting tachycardia indicates the likelihood of parasympathetic cardiovagal dysfunction (the vagus nerve acts like a brake on the heart; a lack of vagal innervation produces a tachycardia). The blood pressure and heart rate are evaluated by measuring both in the lying then standing positions. A failure of the heart rate to increase is another indication of vagal dysfunction. A drop of 20 mm Hg systolic and/or 10 mm Hg diastolic pressures is evidence for likely sympathetic vasoconstrictor abnormality. Sluggish pupillary responses to light and bone-dry hands and feet are other features that implicate autonomic dysfunction. None of these findings is a perfect indicator that the bowel, bladder or erectile difficulty is due to damage to the peripheral autonomic nerves, but they provide a strong inference that this is the case.

When focal pelvic nerve damage is the cause of bladder, bowel or erectile dysfunction, there will always be a history of pelvic or prostatic surgery, with the symptoms starting afterwards. The neurological examination is seldom helpful in these patients.

Investigations. Investigations can be considered in three categories:

1. Those directed at detecting or confirming the presence of a polyneuropathy affecting the somatic peripheral nerves.
2. Tests of other aspects of ANS function.
3. Tests of the function and nerve supply of the distal bowel, bladder, and erectile tissues.

To evaluate a polyneuropathy involving somatic peripheral nerves, the standard approach is to perform nerve conduction studies, often supplemented by electromyographic (EMG) examination of distal limb muscles. Thermal thresholds measured on the feet are also valuable as these assess the function of the small diameter nerve fibers (nerve conduction and EMG studies evaluate the large diameter fibers).

Another approach to attempting to establish that bladder, bowel or erectile dysfunction are being caused by dysautonomia, is to test other parts of the ANS — as one does clinically (described above). Thus, heart rate responses to stimuli, such as deep breathing, or quantitative measures of blood pressure changes to standing or elevated on a tilt table, can be measured to detect cardiovascular autonomic dysfunction [22]. A variety of tests for sweating, including sympathetic skin responses, can be used to assess those autonomic peripheral nerve fibers [23]. If such abnormalities are found, then by inference, the bladder, bowel or erectile dysfunction is probably on the basis of autonomic nerve damage. However, it must be emphasized that this is an "apples and oranges" situation, i.e. one is testing the apples and making inferences about the state of the oranges. The potential pitfalls are evident.

Specific tests of bladder, bowel and erectile function are described in Chapters 1, 2, 3 and 4.

In *summary*, laboratory testing to prove a neurogenic cause for bladder, bowel or erectile dysfunction in the context of polyneuropathies requires particular expertise and has significant limitations. It is often more appropriate to make the diagnosis on clinical grounds and to treat appropriately, without extensive testing.

TREATMENT

The approach to treatment is no different to other neurological causes of these symptoms. Such treatments are discussed in Chapter 2 (urinary incontinence and retention), Chapter 3 (constipation and fecal incontinence) and Chapter 4 (male and female sexual dysfunction).

KEYPOINTS:

- Laboratory testing to prove a neurogenic cause for bladder, bowel or erectile dysfunction in the context of polyneuropathies requires particular expertise and has significant limitations. It is often more appropriate to make the diagnosis on clinical grounds and to treat appropriately, without extensive testing.

CASE-ORIENTED MULTIPLE CHOICE QUESTIONS

▶ Which of the following statements regarding diabetic autonomic neuropathy (DAN) is correct?

A. It is usually present in patients with marked polyneuropathy.
B. It usually presents with abdominal bloating due to gastric atony.
C. A common early symptom is dry hands and feet due to impaired sweating.
D. It is essentially a benign disorder producing trivial symptoms.
E. It occurs only in type I diabetics.

The answer is A. Most, but not all, patients with DAN have marked polyneuropathy. The presentation of DAN is quite variable, so bloating or sweating abnormalities are just two of several possible presenting symptoms. Both types of diabetics may develop DAN, and it is associated with higher mortality compared with patients without DAN.

▶ Which of the following statements regarding erectile dysfunction (ED) in diabetic men is correct?

A. This is a rare condition.
B. When it does occur, the patient usually has clear evidence of severe polyneuropathy.
C. When it does occur, the patient usually has many other manifestations of DAN.
D. It is thought to be usually psychogenic in origin.
E. One way of treating this is with intracavernosal injection therapy.

The answer is E. Unfortunately, ED is common in diabetic men, and may occur in the absence of significant polyneuropathy and other manifestations of DAN. Although it may be psychogenic in origin, as in any man with ED, it is thought to be mainly due to damage to the autonomic nerves supplying the corpora cavernosae. Treatments include sildenafil, intracorporeal injections of vasoactive medications, and penile implants.

▶ Which one of the following statements is correct regarding diabetic cystopathy?

A. It is the leading cause of chronic renal failure in diabetics.
B. It is an acute syndrome consisting of acute urinary retention.
C. It is usually associated with a small spastic bladder.
D. It predisposes to urinary tract infections.
E. Because of the risk of infection, this disorder should never be treated with intermittent catheterization.

The answer is D. Urinary infections are a common consequence. Although intermittent catheterization can cause bladder infections, this risk is low when it is done correctly, and some patients with diabetic cystopathy are best managed in this way. Chronic renal failure is usually caused by glomerular damage rather than recurrent infections. The bladder is often large and "flabby", but can also be small and fibrotic.

▶ Amyloidosis affects the peripheral somatic and autonomic nervous systems. Which of the following statements is correct?

A. It is a common disorder.
B. It is often hereditary.
C. It is a benign condition in which the life expectancy is normal.

D. Symptoms from the involvement of the peripheral somatic nerves are usually incon spicuous, consisting of mild foot numbness.
E. Autonomic manifestations are confined to bladder and bowel dysfunction, and ED (in male patients).

The answer is B. This is a rare and most often hereditary disorder that carries a poor prognosis. Polyneuropathy symptoms can be marked, and the dysautonomia widespread, with orthostatic hypotension being a prominent feature.

▶ Of the inflammatory neuropathies — Guillain-Barré syndrome (GBS) and chronic inflammatory demyelinating neuropathy (CIDP), which of the following is correct?

A. Autonomic dysfunction is common in GBS.
B. Autonomic dysfunction is common in CIDP.
C. Most patients with GBS require catheterization of the bladder because nearly all have severe bladder atony from involvement of the autonomic innervation to the bladder.
D. Cardiac dysrhythmias occur frequently in GBS, but are essentially benign.
E. Orthostatic hypotension is an inevitable and serious complication in CIDP.

The answer is A. Autonomic dysfunction is common in GBS, but not CIDP. Bladder catheterization is usually done for ease of bladder management, rather than because of bladder dysfunction per se. Patients with GBS sometimes have acute and fatal or potentially fatal cardiac dysrhythmias.

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