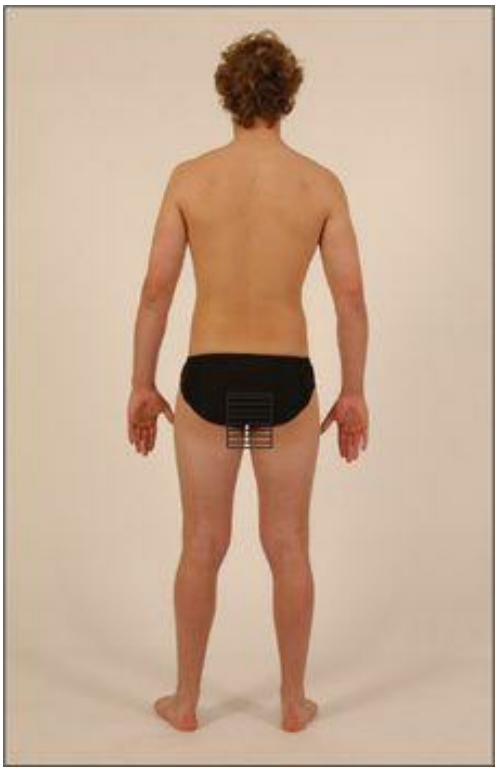


Figure 11.100 Conus medullaris or cauda equina lesion—saddle anaesthesia

TABLE 11.30 Important motor and reflex changes of spinal cord compression

See Figures 11.98 to 11.100 for sensory changes.
Upper cervical
Upper motor neurone signs in the upper and lower limbs.
C5
• Lower motor neurone weakness and wasting of rhomboids, deltoids, biceps and brachioradialis.
• Upper motor neurone signs affect the rest of the upper and all the lower limbs. The biceps jerk is lost. The brachioradialis jerk is inverted.
C8
• Lower motor neurone weakness and wasting of the intrinsic muscles of the hand.
• Upper motor neurone signs in the lower limbs.

Midthoracic
Intercostal paralysis.
Upper motor neurone signs in the lower limbs.
Loss of upper abdominal reflexes at T7 and T8.
T10–T11
• Loss of the lower abdominal reflexes and upward displacement of the umbilicus.
• Upper motor neurone signs in the lower limbs.
L1
• Cremasteric reflex is lost (normal abdominal reflexes).
• Upper motor neurone signs in the lower limbs.
L4
• Lower motor neurone weakness and wasting of the quadriceps.
• Knee jerks lost.
• Ankle jerks may be hyperreflexic with extensor plantar response (upgoing toes), but more often the whole conus is involved, causing a lower motor neurone lesion.
L5–S1
• Lower motor neurone weakness of knee flexion and hip extension (S1) and abduction (L5) plus calf and foot muscles.
• Knee jerks present.
• No ankle jerks or plantar responses.
• Anal reflex present.
S3–S4
• No anal reflex.
• Saddle sensory loss.
• Normal lower limbs.
Causes of spinal cord compression
1 Vertebral
• Spondylosis

• Spondylosis
• Trauma
• Prolapse of a disc
• Tumour
• Infection
2 Outside the dura
• Lymphoma, metastases
• Infection—e.g. abscess
3 Within the dura but extramedullary
• Tumour—e.g. meningioma, neurofibroma
4 Intramedullary*
• Tumour—e.g. glioma, ependymoma
• Syringomyelia
• Haematomyelia

* Lower motor neurone signs may extend for several segments, and spastic paralysis occurs late, unlike the situation with extramedullary lesions.

After carefully examining the lower limbs (see above) determine the level of any sensory impairment (Table 11.31). Then examine the back for signs of a local lesion. Look for deformity, scars and neurofibromas. Palpate for vertebral tenderness and auscultate down the spine for bruits. Next examine the upper limbs and cranial nerves to determine the upper level if this is not already obvious.

TABLE 11.31 Important patterns of abnormal sensation

Sign	Location of lesion
Total unilateral loss of all forms of sensation	Thalamus or upper brainstem (extensive lesion)
Pain and temperature loss on one side of face and opposite side of body	Medulla involving descending nucleus of spinal tract of the fifth nerve and ascending spinothalamic tract (lateral medullary lesion) (Figure 11.102)
Bilateral loss of all forms of sensation below a definite level	Spinal cord lesion (if only pain and temperature affected: anterior cord lesion)
Unilateral loss of pain and temperature below a definite level	Partial unilateral spinal cord lesion on opposite side (Brown-Séquard syndrome) (Figure 11.101)
Loss of pain and temperature over several segments but normal sensation above and below	Intrinsic spinal cord lesion near its centre anteriorly (involves the crossing fibres), e.g. syringomyelia, intrinsic cord tumour (Note: more posterior lesions cause proprioceptive loss)
Loss of sensation over many segments with sacral sparing	Intrinsic cord compression more likely
Saddle sensory loss (lowest sacral segments)	Cauda equina lesion (touch preserved in conus medullaris lesions)

Loss of position and vibration sense only	Posterior column lesion
Glove and stocking loss (hands and feet)	Peripheral neuropathy
Loss of all forms of sensation over a well-defined body part only	Posterior root lesion (purely sensory) or peripheral nerve (often motor abnormality associated)

Important spinal cord syndromes

Brown-Séquard syndrome[aaa]

Clinical features are shown in Figure 11.101. These signs result from hemisection of the cord.

• **Motor changes:** (i) upper motor neurone signs below the hemisection on the same side as the lesion; (ii) lower motor neurone signs at the level of the hemisection on the same side.

• **Sensory changes:** (i) pain and temperature loss on the opposite side to the lesion—note that the upper level of sensory loss is usually a few segments below the level of the lesion; (ii) vibration and proprioception loss occur on the same side; (iii) detection of light touch is often normal.

• **Causes:** (i) multiple sclerosis; (ii) angioma; (iii) trauma; (iv) myelitis; (v) post-radiation myelopathy.

Figure 11.101 Brown-Séquard syndrome
Loss of pain and temperature on the opposite side to the lesion, with loss of vibration and proprioception on the same side as the lesion.

Subacute combined degeneration of the cord (vitamin B_{12} deficiency)

Clinical features are (i) posterior column loss symmetrically (vibration and joint position sense), causing an ataxic gait; (ii) upper motor neurone signs in the lower limbs symmetrically with absent ankle reflexes; knee reflexes may be absent or, more often, exaggerated. There may also be (iii) peripheral sensory neuropathy (less common and mild); (iv) optic atrophy; (v) dementia.

Dissociated sensory loss

This usually indicates spinal cord disease but may occur with a peripheral neuropathy.

- **Causes of spinothalamic loss only:** (i) syringomyelia; (ii) Brown-Séquard syndrome (contralateral leg); (iii) anterior spinal artery thrombosis; (iv) lateral medullary syndrome (contralateral to the other signs) (Figure 11.102); (v) small fibre peripheral neuropathy (e.g. diabetes mellitus, amyloid).

- **Causes of dorsal column loss only:** (i) subacute combined degeneration; (ii) Brown-Séquard syndrome (ipsilateral leg); (iii) spinocerebellar degeneration (e.g. Friedreich's[bbb] ataxia); (iv) multiple sclerosis; (v) tabes dorsalis; (vi) peripheral neuropathy (e.g. diabetes mellitus, hypothyroidism); (vii) sensory neuronopathy (a dorsal root ganglionopathy which may be caused by carcinoma, diabetes mellitus or Sjögren's

which may be caused by carcinoma, diabetes mellitus or Sjögren's syndrome).

Figure 11.102 Pattern of sensory loss in the lateral medullary syndrome

Syringomyelia (a central cavity in the spinal cord)

• **Clinical triad:** (i) loss of pain and temperature over the neck, shoulders and arms (a 'cape' distribution); (ii) amyotrophy (atrophy and areflexia) of the arms; (iii) upper motor neurone signs in the lower limbs.

There may also be thoracic scoliosis due to asymmetrical weakness of the paravertebral muscles.

An extensor plantar response plus absent knee and ankle jerks

• **Causes:** (i) subacute combined degeneration of the cord (B_{12} deficiency); (ii) conus medullaris lesion; (iii) combination of an upper motor neurone lesion with cauda equina compression or peripheral neuropathy, such as a stroke in a diabetic; (iv) syphilis (taboparesis); (v) Friedreich's ataxia; (vi) motor neurone disease; (vii) human T-cell lymphotropic virus (HTLV-I)

infection.

A summary of the features that differentiate intramedullary from extramedullary cord lesions is presented in Table 11.32.

TABLE 11.32 Differentiating intramedullary from extramedullary cord lesions

Intramedullary	Extramedullary
Root pains rare	Root pains common
Late onset of corticospinal signs	Early onset of corticospinal signs
Lower motor neurone signs extend for several segments	Lower motor neurone signs localised
Dissociated sensory loss (pain and temperature) may be present	Brown-Séquard syndrome if lateral cord compression
Normal or minimally altered cerebrospinal fluid findings	Early, marked cerebrospinal fluid abnormalities
May have sacral sparing	

Myopathy

Muscle weakness can be due to individual peripheral nerve lesions, mononeuritis multiplex, peripheral neuropathy or spinal cord disease. Each of

these has a characteristic pattern. Primary disease of muscle (myopathy) also causes weakness. There is no sensory loss with myopathy, which is an important clue. The motor weakness is similar to that of the lower motor neurone type. There are two major patterns: proximal myopathy and distal myopathy.

Proximal myopathy is the more common form. On examination there is proximal muscle wasting and weakness (Tables 11.33 and 11.34; Figures 11.103 and 11.104). Reflexes involving these muscles may be reduced. This can be caused by genetic (e.g. muscular dystrophy) or acquired disease. *Distal myopathy* also occurs and is always genetic, although peripheral neuropathy is a much more common cause of distal muscle weakness. If the distal limbs are affected, consider hereditary motor and sensory neuropathy (Table 11.35). Motor neurone disease also causes weakness without any sensory loss.

TABLE 11.33 Causes (differential diagnosis) of proximal weakness and myopathy

Causes of proximal weakness
Myopathy (q.v.)
Neuromuscular junction disease, e.g. myasthenia gravis
Neurogenic, e.g. motor neurone disease, polyradiculopathy, Kugelberg-Welander disease (proximal muscle wasting and

fasciculation due to anterior horn cell disease—autosomal recessive)
Causes of myopathy
Hereditary muscular dystrophy (Table 11.34)
Congenital myopathies (rare)
Acquired myopathy (mnemonic, PACE, PODS):
Polymyositis or dermatomyositis (Figure 11.104)
Alcohol, AIDS (HIV infection)
Carcinoma
Endocrine—e.g. hyperthyroidism, hypothyroidism, Cushing's syndrome, acromegaly, hypopituitarism
Periodic paralysis (hyperkalaemic, hypokalaemic or normokalaemic)
Osteomalacia
Drugs—e.g. clofibrate, chloroquine, steroids, zidovudine
Sarcoidosis
Note: Causes of *proximal myopathy with a peripheral neuropathy* include:
Paraneoplastic syndrome
Alcohol
Hypothyroidism
Connective tissue diseases

TABLE 11.34 Muscular dystrophies

1 Duchenne's* (pseudohypertrophic)

• Affects only males (sex-linked recessive) • Calves and deltoids: hypertrophied early, weak later • Proximal weakness: early • Dilated cardiomyopathy
2 Becker[†] • Affects only males (sex-linked recessive) • Similar clinical features to Duchenne's except for less heart disease, a later onset and less-rapid progression
3 Limb girdle • Males or females (autosomal recessive), onset in the third decade • Shoulder or pelvic girdle affected • Face and heart usually spared
4 Facioscapulohumeral • Males or females (autosomal dominant) • Facial and pectoral weakness with hypertrophy of deltoids
5 Dystrophia myotonica (autosomal dominant)

[*] Guillaume Duchenne (1806–75), brilliant eccentric who founded French neurology. He died of a stroke.

[†] Peter Becker (b. 1908), German professor of genetics.

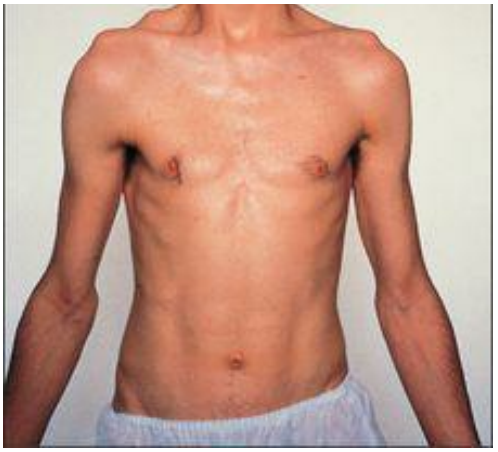


Figure 11.103 Fascioscapular muscular dystrophy

From Mir MA, Atlas of Clinical Diagnosis, 2nd edn. Edinburgh: Saunders, 2003.

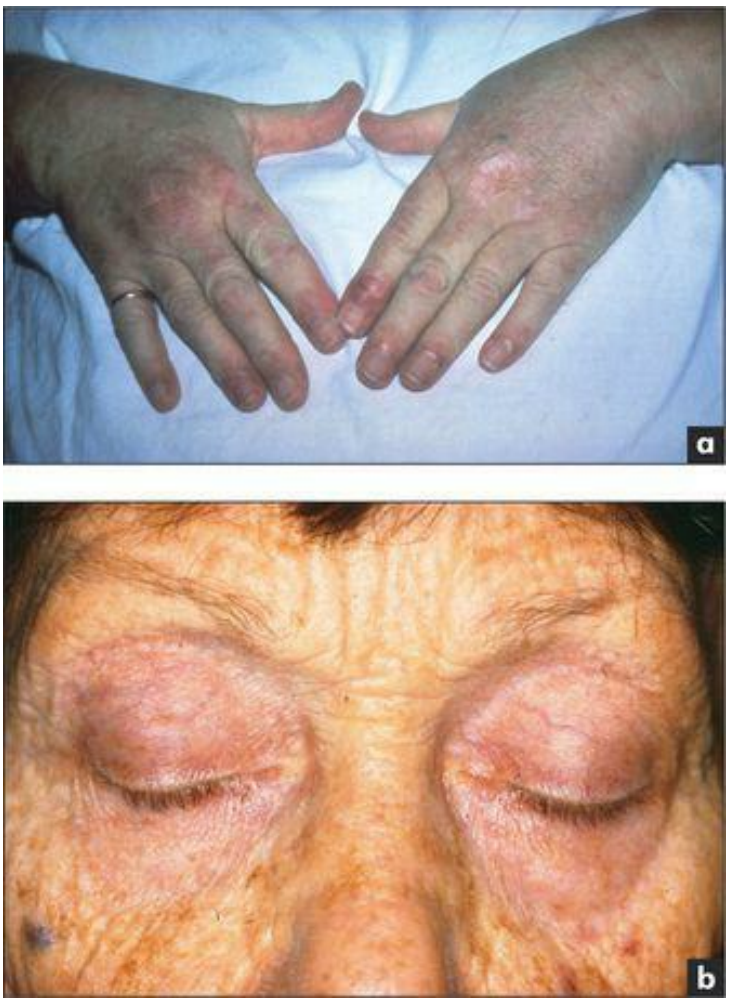


Figure 11.104 Dermatomyositis
(a) Gottren's sign in dermatomyositis—heliotrope (lilac-coloured) flat topped papules, which occur over the knuckles, but may also be seen over the elbows or knees and may ulcerate. (b) Dermatomyositis may also cause a heliotrope rash on the face (especially on the eyelids, upper cheeks and forehead), periorbital oedema, erythema, maculopapular eruptions and

scaling dermatitis.
Dermatomyositis and the closely related condition polymyositis are idiopathic myopathies. Up to 10% of adult patients with dermatomyositis may have an underlying malignancy.

From McDonald FS, ed., Mayo Clinic images in internal medicine, with permission. ©Mayo Clinic Scientific Press and CRC Press.

TABLE 11.35 Features of hereditary motor and sensory neuropathy (Charcot-Marie-Tooth* disease)

1 Pes cavus (short-arched feet)
2 Distal muscle atrophy due to peripheral nerve degeneration. This does not usually extend above the elbows or above the middle third of the thighs. Peroneal muscle atrophy gives the legs the shape of an inverted champagne bottle
3 Absent reflexes
4 Slight or no sensory loss in the limbs
5 Thickened nerves
6 Optic atrophy, Argyll Robertson pupils (rare)

* Jean Martin Charcot (1825–93), Parisian physician and neurologist; Pierre Marie (1853–1940), Parisian neurologist and Charcot's greatest pupil; and Howard Henry Tooth (1856–1926), London physician, who described the condition independently in 1886. Type I is usually autosomal dominant.

Dystrophia myotonica

If this disease (which is inherited as an autosomal-dominant condition) is suspected because of an inability on the part of the patient to let go when shaking hands (myotonia), or because general inspection reveals the characteristic appearance (Figure 11.105), examine as follows.

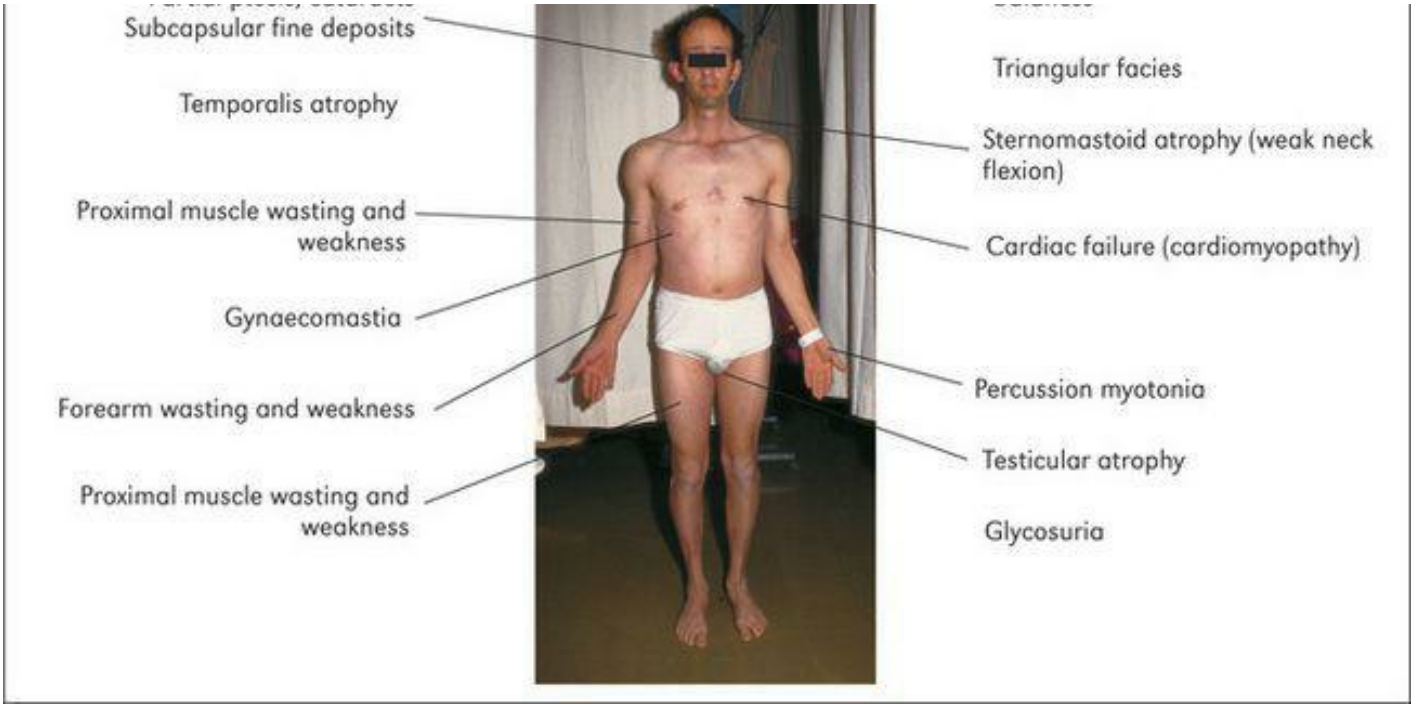


Figure 11.105 Dystrophia myotonica

Observe the face for frontal baldness (the patient may be wearing a wig), the expressionless triangular facies, atrophy of the temporalis muscle and partial ptosis. Thick spectacles, a traditional sign of this disease, are not often seen now because of lens replacement surgery. The eyes should still be examined, as these patients can develop characteristic iridescent cataracts and subcapsular fine deposits.

Look at the neck for atrophy of the sternocleidomastoid muscles and then test neck flexion (neck flexion is weak while extension is normal).

Go to the upper limbs. Shake hands and test for percussion myotonia. Tapping over the thenar eminence causes contraction and then slow relaxation of abductor pollicis brevis. Examine the arm now for signs of wasting and weakness distally (forearms are usually affected first) and proximally. There are no sensory changes.

Go to the chest and look for gynaecomastia (uncommon). Examine the cardiovascular system for cardiomyopathy. Next palpate the testes for atrophy. Examine the lower limbs. The tibial nerves are affected first. Always ask to test the urine for sugar (diabetes mellitus is associated with this disease).

Note: Muscle myotonia can also occur in the hereditary diseases myotonia congenita (autosomal dominant or recessive) and hereditary paramyotonia (autosomal dominant cold-induced myotonia).

Myasthenia gravis

Myasthenia gravis is an autoimmune disease of the neuromuscular junction. There are circulating antibodies against acetylcholine receptors. It differs from the proximal myopathies in that muscle power decreases with use. There is little muscle wasting and no sensory change.

It is necessary to test for muscle fatigue. Test the oculomotor muscles by asking the patient to sustain an upward gaze by looking up at the ceiling for 1 minute, and watch for progressive ptosis (Figure 11.106). Test the Peek sign for orbicularis oculi weakness. Ask the patient to close the eyes; if positive, within 30 seconds the lid margin will begin to separate, showing the sclera. This test strongly increases the likelihood of myasthenia (positive LR = 30.0).[22] Then test the proximal limb girdle muscles—ask the patient to hold the arms above the head. The examiner can repeatedly press the abducted arms down until they weaken. Power will decrease with repeated muscle contraction.

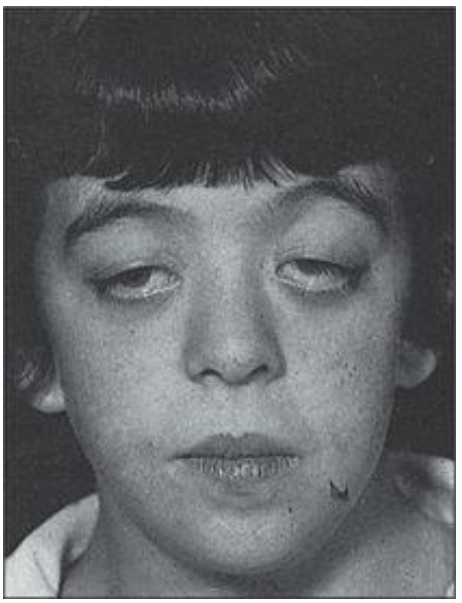


Figure 11.106 Bilateral ptosis after upward gaze in myasthenia gravis

Look for a thymectomy scar (over the sternum)—thymectomy is often undertaken as treatment for generalised myasthenia.

The cerebellum (Figure 11.107)

If the patient complains of clumsiness or problems with coordination of movement, a cerebellar examination is indicated. Signs of cerebellar disease occur on the same side as the lesion in the brain. This is because most cerebellar fibres cross twice in the brainstem, both on entry to and exit from the cerebellum. Proceed as follows with the examination

the cerebellum. Proceed as follows with the examination:

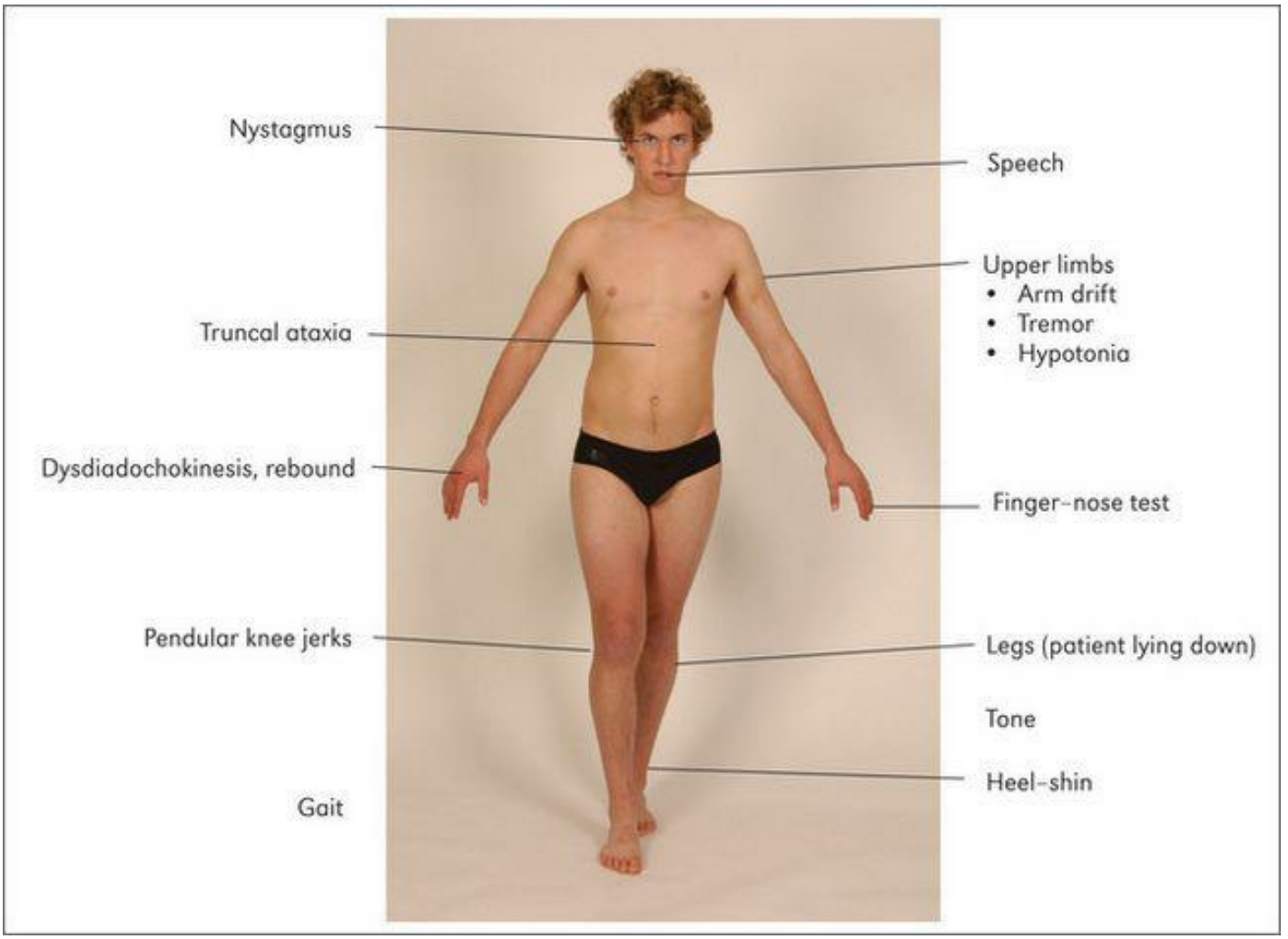


Figure 11.107 The cerebellar examination

Look first for nystagmus—usually jerky horizontal nystagmus with an increased amplitude on looking towards the side of the lesion. The direction of fast movement is the side of the lesion. Assess speech next. Ask the patient to say 'British Constitution' or 'West Register Street' (Figure 11.108). Cerebellar speech is jerky, explosive and loud, with an irregular separation of syllables.

Figure 11.108 West Register Street, Edinburgh

Go to the upper limbs. Ask the patient to extend the arms and look for upward arm drift due to hypotonia of the agonist muscles. Test tone. Hypotonia[ccc] is due to loss of a facilitatory influence on the spinal motor neurones.

Next perform the finger–nose test. The patient touches the nose, then rotates the finger and touches the examiner's finger. Note any intention tremor (tremor which increases as the target is approached—this is due to loss of cerebellar connections in the brainstem) and past-pointing (the patient overshoots the target). Test rapidly alternating movements: the patient taps alternately the palm and back of one hand on the other hand or thigh. Inability to perform this movement smoothly is called dysdiadochokinesis. Now test rebound—ask the patient to lift the arms quickly from the sides, then stop (incoordination of antagonist and agonist action causes the patient to be unable to stop the arms). Before testing, always demonstrate these movements for the patient's benefit.

Go on to examine the legs. Again, test tone here. Then perform the heel–shin test looking for accuracy of fine movement when the patient slides the heel down the shin slowly on each side for several cycles. Then ask the patient to lift the big toe up to touch the examiner's finger, and look for intention tremor and past-pointing. Ask the patient then to tap each heel on the other shin.

Look for truncal ataxia by asking the patient to fold the arms and sit up. While the patient is sitting, ask him or her to put the legs over the side of the bed and test for pendular knee jerks (the lower leg continues to swing a number of times before coming to rest—this is evidence of hypotonia).

Test gait (the patient will stagger towards the affected side if there is a unilateral cerebellar hemisphere lesion).

If there is an obvious unilateral cerebellar problem examine the cranial

If there is an obvious unilateral cerebellar problem examine the cranial nerves for evidence of a cerebellopontine angle tumour (fifth, seventh and eighth nerves affected) or the lateral medullary syndrome, and auscultate over the cerebellum. Always look in the fundi for papilloedema. Next examine for peripheral signs of malignant disease and for vascular disease (carotid or vertebral bruits). Examine the base of the skull for scars from previous neurosurgery.

If there is evidence of a midline lesion, such as truncal ataxia, abnormal heel–toe walking or abnormal speech, consider either a midline tumour or a paraneoplastic syndrome (Table 11.36). If there is bilateral disease, look for signs of multiple sclerosis, Friedreich's ataxia (pes cavus is the most helpful initial clue) (Table 11.37) and hypothyroidism (rare). Alcoholic cerebellar degeneration (which affects the anterior lobe of the cerebellar vermis) classically spares the arms. If there are, in addition, upper motor neurone signs, consider the causes in Table 11.38.

TABLE 11.36 Causes of cerebellar disease

Rostral vermis lesion (only lower limbs affected)
Usually due to alcohol
Unilateral
1 Space-occupying lesion (tumour, abscess, granuloma)
2 Ischaemia (vertebrobasilar disease)

3 Multiple sclerosis
4 Trauma
Bilateral
1 Drugs—e.g. phenytoin
2 Alcohol (both acutely and chronically, possibly due to thiamine deficiency)
3 Friedreich's ataxia
4 Hypothyroidism
5 Paraneoplastic syndrome
6 Multiple sclerosis
7 Trauma ('punch drunk')
8 Arnold-Chiari malformation
9 Large space-occupying lesion, cerebrovascular disease
Midline
1 Paraneoplastic syndrome
2 Midline tumour

TABLE 11.37 Clinical features of Friedreich's ataxia (autosomal recessive)

This is usually a young person with:
1 Cerebellar signs (bilateral) including nystagmus
2 Pes cavus.* Cocking of the toes. Kyphoscoliosis
3 Upper motor neurone signs in the limbs (although reflexes are

absent)
4 Peripheral neuropathy
5 Posterior column loss in the limbs
6 Cardiomyopathy (ECG abnormalities occur in more than 50% of cases)
7 Diabetes mellitus (common)
8 Optic atrophy (uncommon)
9 Normal mentation

* Other causes of pes cavus include hereditary motor and sensory neuropathy, spinocerebellar degeneration or neuropathies in childhood.

TABLE 11.38 Causes of spastic and ataxic paraparesis (upper motor neurone and cerebellar signs combined)

In adolescence
Spinocerebellar degeneration—e.g. Marie's spastic ataxia
In young adults
Multiple sclerosis
Spinocerebellar degeneration
Syphilitic meningomyelitis
Arnold-Chiari malformation or other lesion at the craniospinal junction
In later life

Multiple sclerosis
Syringomyelia
Infarction (in upper pons or internal capsule on one side—'ataxic hemiparesis')
Lesion at the craniospinal junction—e.g. meningioma
Note: Unrelated diseases that are relatively common (e.g. cervical spondylosis and cerebellar degeneration from alcohol) may cause a similar clinical picture.

Remember that there are important reciprocal connections between the cerebellum and the parietal and frontal lobes. These explain the problems cerebellar abnormalities can cause with functions other than coordination. Loss of verbal fluency, grammatical problems with speech, difficulty with memory and planning can all sometimes be features of cerebellar disease.

Parkinson's disease

This is a common extrapyramidal disease of middle to old age (1% of people older than 65) where there is degeneration of the substantia nigra and its pathways. This results in dopamine deficiency and a relative excess of cholinergic transmission in the caudate nucleus and putamen, which causes excessive supraspinal excitatory drive. There may be a history of insidious and asymmetrical onset. Non-specific symptoms (sleep abnormalities, constipation, depression and dementia) may precede or accompany the classic tremor.

Examine as follows.[23]

Inspection

Note the lack of facial expression, which leads to a mask-like facies. The posture is characteristically flexed and there are few spontaneous movements.

Gait and movements

Ask the patient to rise from a chair, walk, turn quickly, stop and start.

The characteristic gait is described as *shuffling*—there are small steps,

and the patient hardly raises the feet from the ground. There is often difficulty in initiating walking, but once it begins the patient hurries (festination) and has difficulty stopping. The Parkinsonian patient seems always to be trying to catch up with the centre of gravity. There is a lack of the normal arm swing. Walking heel-to-toe will be difficult.

Testing for *propulsion* or *retropulsion* (propulsion involves pushing the patient from behind and retropulsion pushing from in front) is of uncertain value and must be done with some caution because the patient may be unable to stop and may fall over. The examiner can stand behind the patient and pull him or her backwards, but should stand braced to catch the patient.

Bradykinesia (a decrease in the speed and amplitude of complex movements) may be the result of a lesion in the nigrostriatal pathway (a dopaminergic pathway), which affects connections between the caudate nucleus, putamen and motor cortex, causing abnormal movement programming and abnormal recruitment of single motor units. Two simple tests (Figure 11.109) for this are *finger tapping* and *twiddling*. Ask the patient to tap the fingers in turn onto a surface repeatedly, quickly and with both hands at once. Twiddling is rotating the hands around each other in front of the body. These movements are slow and clumsy in Parkinsonian patients but obviously depend on motor and cerebellar function as well. Difficulty getting out of a chair can be another sign of bradykinesia and patients often have difficulty turning over in bed.

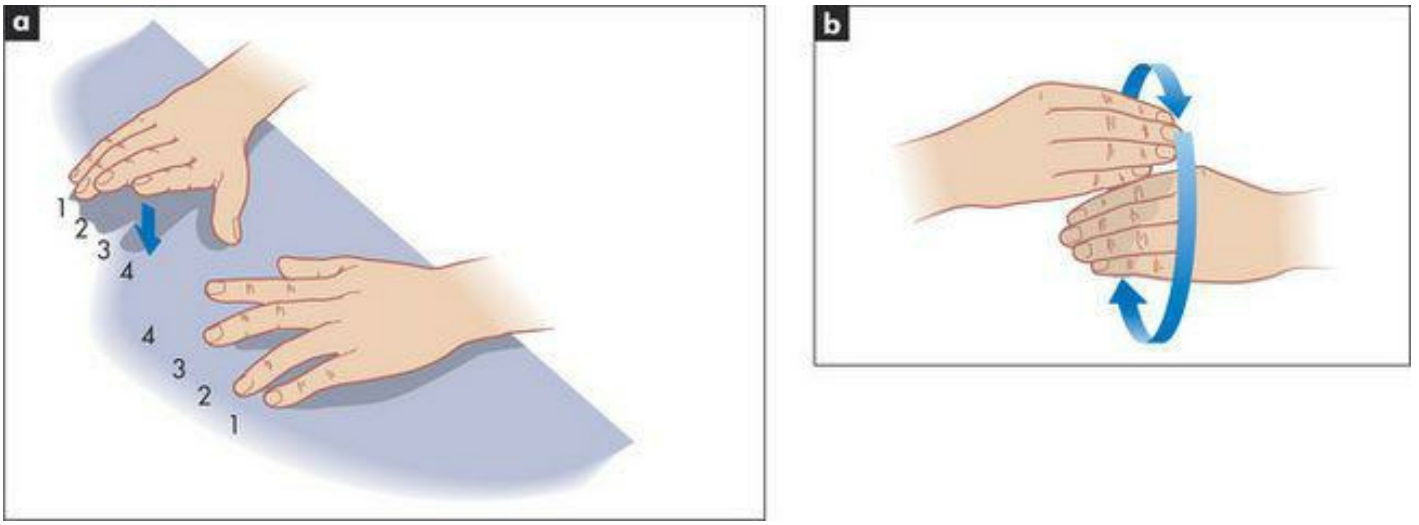


Figure 11.109 Detecting bradykinesia
(a) Tapping the fingers. (b) Twiddling.

Kinesia paradoxica is the striking ability of a patient to perform rapid movements (especially if startled) but not slow ones; for example, the patient may be able to run down the stairs in response to a fire alarm but be unable to stop at the bottom—this is *not* a recommended test.

Tremor

Have the patient return to bed. Look for a resting tremor, which is often asymmetrical. The characteristic movement is described as pill-rolling. Movement of the fingers at the metacarpophalangeal joints is combined with the movements of the thumb. Various attending movements may also occur at the wrist. On finger–nose testing the resting tremor decreases, but a faster action tremor may supervene.

Tremor can be facilitated by getting the patient to perform 'serial 7s'—take 7 from 100, then 7 from the answer and so forth (mental stimulation)—or to move the contralateral limb (e.g. by rapidly opposing the contralateral thumb and fingers). Other types of tremor are summarised in Table 11.39.

TABLE 11.39 A classification of non-physiological tremor*

1 Parkinsonian—resting tremor
2 Postural/action tremor; present throughout movement; the following most often cause a static or postural tremor of the outstretched arms:
(a) idiopathic (most common)
(b) anxiety
(c) drugs
(d) familial
(e) thyrotoxicosis
3 Essential/familial
4 Intention tremor (cerebellar disease); increases towards the target

5 Midbrain ('red nucleus') tremor—abduction-adduction movements of upper limbs with flexion-extension of wrists (usually associated with intention tremor)

NB: Flapping (asterixis) is not strictly a tremor but a sudden brief loss of tone in hepatic failure, cardiac failure, respiratory failure or renal failure (metabolic encephalopathies).

* Tremor is a rhythmic oscillation of a part of the body around a fixed point.

Tone

Test tone at both wrists. The characteristic increase in tone is called cogwheel or plastic (lead pipe) rigidity. Tone is increased with an interrupted nature, the muscles giving way with a series of jerks. If hypertonia is not obvious, obtain reinforcement by asking the patient to turn the head from side to side or to wave the contralateral arm. Cogwheel rigidity occurs because the exaggerated stretch reflex is interrupted by tremor.

Remember, the signs are often asymmetrical early in the course of Parkinson's disease.

Face

There may be *titubation* (tremor) of the head, absence of blinking, dribbling of saliva and lack of facial expression. Test the *glabellar tap* (reflex)—keeping your finger out of the patient's line of vision, tap the middle of the forehead (glabella) with your middle finger (Figure 11.110). This sign is positive when the patient continues to blink as long as the examiner taps. Normal people blink only a couple of times and then stop. The glabellar reflex is a primitive reflex which is also frequently present in frontal lobe disease.

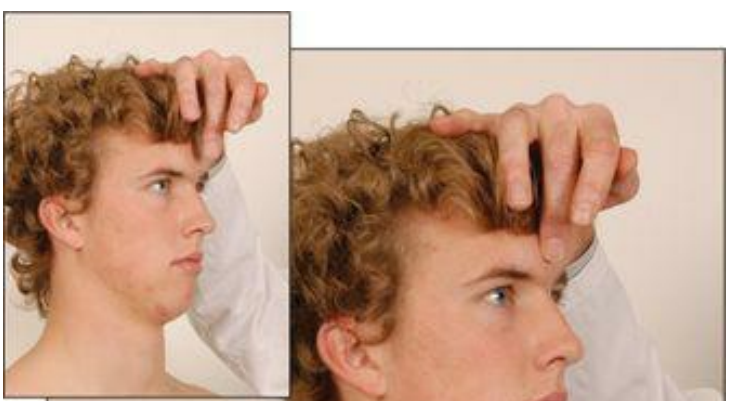


Figure 11.110 The glabellar tap (Wilson's sign)

Assess *speech*, which is typically monotonous, soft and faint, lacking intonation. Sometimes palilalia is present; this is repetition of the end of a word (the opposite of stuttering).

Now test the ocular movements, particularly for *weakness of upward gaze*. Isolated failure of upward gaze is a feature of Parkinson's disease. There is a separate group of patients with marked rigidity and paralysis of gaze who should be diagnosed as having progressive supranuclear palsy rather than Parkinson's disease. These people develop loss first of downward gaze, then of upward gaze and finally of horizontal gaze.

Feel the brow for greasiness (seborrhoea) or sweatiness, due to associated autonomic dysfunction. Orthostatic hypotension may also be present for the same reason.

The palmomental reflex is commonly present in these patients and tends to be more prominent in those with severe akinesia. Dementia develops in 30% of patients.

Writing

Ask the patient to write his or her name and address. *Micrographia* (small writing) is characteristic. The patient may also be unable to do this because of the development of dementia, a late manifestation. Test the higher centres if appropriate. See *Good signs guide 11.3*.

GOOD SIGNS GUIDE 11.3 Parkinson's disease

Sign	Positive LR	Negative LR
Rigidity, tremor and bradykinesia all present	2.2	0.5
Difficulty walking (heel–toe)	2.9	0.32
Asymmetrical onset, no atypical features and no alternative diagnosis	4.1	0.4
Glabellar tap	4.5	0.13

From McGee S, Evidence-based physical diagnosis, 2nd edn. St Louis: Saunders, 2007.

Causes of Parkinson's syndrome

These are shown in Table 11.40.

TABLE 11.40 Causes of Parkinson's syndrome

Idiopathic: Parkinson's disease
Drugs: e.g. phenothiazines, methyldopa
Post-encephalitis (now very rare)
Other: toxins (carbon monoxide, manganese), Wilson's disease, progressive supranuclear palsy, Steele-Richardson syndrome, Shy-Drager syndrome, syphilis, tumour (e.g. giant frontal meningioma)
Atherosclerosis is a controversial cause

Other extrapyramidal movement disorders (dyskinesia)

Chorea

Here there is a lesion of the corpus striatum, which causes non-repetitive, abrupt, involuntary jerky movements. These may be unilateral or generalised. Often the patient attempts to disguise this by completing the involuntary movements with a voluntary one. In this disease, dopaminergic pathways dominate over cholinergic transmissions.

Chorea can usefully be distinguished from hemiballismus, athetosis and pseudoathetosis. *Hemiballismus* is due to a subthalamic lesion on the side opposite the movement disorder. It causes unilateral wild throwing movements of the proximal joints. There may be skin excoriation due to limb trauma. These movements may persist during sleep. *Athetosis* or *dystonia* is due to a lesion of the outer segment of the putamen and causes slow sinuous writhing distal movements that are present at rest. *Pseudoathetosis* is a description given to athetoid movements in the fingers in patients with severe proprioceptive loss (these are especially prominent when the eyes are shut).

If the patient has chorea, proceed as follows. First shake hands. There may be tremor and dystonia superimposed on lack of sustained hand grip ('milkmaid's grip'). Ask the patient to hold out the hands, then look for a choreic (dystonic) posture. This typically involves finger and thumb hyperextension and wrist flexion.

Go to the face and look at the eyes for exophthalmos (thyrotoxicosis), Kayser-Fleischer rings (Wilson's disease) and conjunctival injection (polycythaemia). Ask the patient to poke out the tongue and note frequent retraction of the tongue (serpentine movements). Look for skin rashes (e.g. systemic lupus erythematosus, vasculitis). If the patient is young, examine the heart for signs of rheumatic fever (Sydenham's[ddd] chorea).

Test the reflexes. The abdominal reflexes are usually brisk, but tendon reflexes are reduced and may be pendular (due to hypotonia).

Assess the higher centres for dementia (Huntington's chorea[eee]).

Causes of chorea are shown in Table 11.41.

TABLE 11.41 Causes of chorea

Drugs: e.g. excess levodopa, phenothiazines, the contraceptive pill, phenytoin
Huntington's disease (autosomal dominant)
Sydenham's chorea (rheumatic fever) and other postinfectious states (both rare)
Senility
Wilson's disease
Kernicterus (rare)
Vasculitis or connective tissue disease—e.g. systemic lupus erythematosus (very rare)
Thyrotoxicosis (very rare)
Polycythaemia or other hyperviscosity syndromes (very rare)
Viral encephalitis (very rare)

Dystonia

The patient manifests an involuntary abnormal posture with excessive co-contraction of antagonist muscles. Dystonia may be focal (e.g. spasmodic torticollis), segmental or generalised. Other forms of movement disorder may be present (e.g. myoclonic dystonia). The acute onset of dystonia is seen most commonly as a side-effect of various drugs (e.g. levodopa, phenothiazines, metoclopramide).

The unconscious patient

The rapid and efficient examination of the unconscious patient is important. The word COMA provides a mnemonic for four major groups of causes of unconsciousness.

C CO_2 narcosis (respiratory failure: uncommon).
O Overdose: for example, tranquillisers, alcohol, salicylates, carbon monoxide, antidepressants.
M Metabolic: for example, hypoglycaemia, diabetic ketoacidosis, uraemia, hypothyroidism, hepatic coma, hypercalcaemia, adrenal failure.
A Apoplexy: for example, head injury, cerebrovascular accident (infarction or haemorrhage), subdural or extradural haematoma, meningitis, encephalitis, epilepsy.

Coma occurs when the reticular formation is damaged by a lesion or metabolic abnormality, or when the cortex is diffusely damaged.

General inspection

Remember A–B–C: ***A****irway*, ***B****reathing* and ***C****irculation*.

Airway and breathing

Look to see if the patient is breathing, as indicated by chest wall movement. If not, urgent attention is required, including clearing the airway and providing ventilation. Note particularly the pattern of breathing (see Table 5.11, page 112). Cheyne-Stokes respiration (which may indicate diencephalic injury, but is not specific), irregular ataxic breathing (Biot's breathing, from an advanced brainstem lesion), and deep rapid respiration (e.g. Kussmaul breathing, secondary to a metabolic acidosis, as in diabetes mellitus) are important signs to look for.

Circulation

Look for signs of shock, dehydration and cyanosis. A typical cherry-red colour occurs rarely in cases of carbon monoxide poisoning. Take the pulse rate and blood pressure.

Posture

Look for signs of trauma. Note any neck hyperextension (from meningism in children or cerebellar tonsillar herniation).

Look for:

1. A *decerebrate* or extensor posture, which may be held spontaneously or occur in response to stimuli, and which suggests severe midbrain disease. The arms are held extended and internally rotated and the legs are extended

The arms are held extended and internally rotated and the legs are extended.

2. A *decorticate* or flexor posture, which suggests a lesion above the brainstem. It can be unilateral or bilateral. There is flexion and internal rotation of the arms and extension of the legs.

Involuntary movements

Recurrent or continuous convulsions, which may be focal or generalised, suggest status epilepticus. Myoclonic jerks can occur after hypoxic injury and as a result of metabolic encephalopathy. Remember that complex partial seizure status epilepticus can cause a reduced level of consciousness without convulsive movements.

Level of consciousness

Tickle the patient's nose with cottonwool and watch for facial movements. This is less likely to harm the patient than the traditional method of firm pressing of knuckles over the sternum to cause pain.

Determine the level of consciousness. *Coma* is unconsciousness with a reduced response to external stimuli. Coma in which the patient responds semi-purposefully is considered light. In deep coma there is no response to any stimuli and no reflexes are present (it is usually due to a brainstem or pontine lesion, though drug overdosage, such as with barbiturates, can be responsible).

Stupor is unconsciousness, but the patient can be aroused. Purposeful movements occur in response to painful stimuli.

Drowsiness resembles normal sleepiness. The patient can be fairly easily roused to normal wakefulness, but when left alone falls asleep again. The Glasgow coma scale (Table 11.42) is used to assess the depth of coma more accurately: record the subscores and total scores.

TABLE 11.42 Glasgow coma scale

Add up the score for 1, 2 and 3. Score of 4 or less = very poor prognosis, score >11 = good prognosis for recovery.			
1 Eyes	Open	Spontaneously	4
		To loud verbal command	3
		To pain	2
	No response		1
2 Best motor response	To verbal command	Obeys	6

	To painful stimuli	Localises pain	5
		Flexion–withdrawal	4
		Abnormal flexion posturing	3
		Extension posturing	2
		No response	1
3 Best verbal response		Oriented	5
		Confused, disoriented	4
		Inappropriate words	3
		Incomprehensible sounds	2
		None	1

The neck

If there is no evidence of neck trauma, assess for neck stiffness and Kernig's sign (for meningitis or subarachnoid haemorrhage).

The head and face

Inspect and palpate for head injuries, including Battle's sign,[fff] bruising behind the ear indicating a fracture of the base of the skull. Look for facial asymmetry (i.e. facial weakness). The paralysed side of the face will be sucked in and out with respiration. A painful stimulus (e.g. pressing the supraorbital notch) may produce grimacing and make facial asymmetry more obvious. Note jaundice (e.g. hepatic coma) or manifestations of myxoedema.

The eyes

Inspect the pupils. Very small pupils (but reactive to light) occur in pontine lesions and with narcotic overdoses. One small pupil occurs in Horner's syndrome (e.g. as part of the lateral medullary syndrome or in hypothalamus injury; see Chapter 13). Two midpoint non-reactive pupils suggest midbrain disease, anoxia or drugs (anticholinergics). One dilated pupil suggests a subdural haematoma, raised intracranial pressure (unilateral tentorial herniation) or a subarachnoid haemorrhage from a posterior communicating artery aneurysm. Widely dilated pupils may occur when increased intracranial pressure and coning cause secondary brainstem haemorrhage, or with anticholinergic drugs.

Conjunctival haemorrhage suggests skull fracture. Look in the fundi for

Conjunctival haemorrhage suggests skull fracture. Look in the fundi for papilloedema, diabetic or hypertensive retinopathy, or subhyaloid haemorrhage. The locked-in syndrome is rare; in a lower brainstem lesion patients are awake but can only control their eye movements.

Look at the position of the eyes. Particular cranial nerve palsies may cause deviation of an eye in various directions. The sixth nerve is particularly vulnerable to damage because of its long intracranial course. Deviation of both eyes to one side in the unconscious patient may be due to a destructive lesion in a cerebral hemisphere, which causes fixed deviation towards the side of the lesion. An irritative (epileptic) focus causes the direction of gaze to be away from the lesion. Upward or downward eye deviation suggests a brainstem problem. Trapping of the globe or extraocular muscles by fracture may also lead to an abnormal eye position or to an abnormal eye movement.

Perform *doll's eye testing* by lifting the patient's eyelids and rolling the head from side to side. When vestibular reflexes are intact (i.e. an intact brainstem), the eyes maintain their fixation as if looking at an object in the distance, but change their position relative to the head. This is the normal 'doll's eye phenomenon'. Brainstem lesions or drugs affecting the brainstem cause the eyes to move with the head, so that fixation is not maintained.

Ears and nostrils

Look for any bleeding or drainage of cerebrospinal fluid (the latter indicating a skull fracture). A watery discharge can be simply tested for glucose. The presence of glucose confirms that it is cerebrospinal fluid.

The tongue and mouth

Trauma may indicate a previous seizure, and corrosion around the mouth may indicate ingestion of a corrosive poison. Gum hyperplasia suggests that the patient may be taking phenytoin for epilepsy. Smell the breath for evidence of alcohol poisoning, diabetic ketosis, hepatic coma or uraemia. Remember that alcohol ingestion may be associated with head injury. Test the gag reflex; its absence may indicate brainstem disease or deep coma, but is not a specific sign. Bite marks on the tongue suggest that an epileptic seizure may have been the cause of unconsciousness.

The upper and lower limbs

Look for injection marks (drug addiction, diabetes mellitus). Test tone in the normal way and by picking up the arm and letting it fall. Compare each side,

assessing for evidence of hemiplegia. In coma and acute cerebral hemiplegia, the muscle stretch reflexes may be normal or reduced at first on the paralysed side. Later the muscle stretch reflexes become increased and the cutaneous reflexes are absent.

Test for pain sensation by placing a pen over a distal finger or toe just below the nail bed. Press firmly and note if there is arm or leg withdrawal. Test all limbs. There will be no response to pain if sensation is absent or if the coma is deep. If sensation is intact but the limb is paralysed, there may be grimacing with movement of the other limbs.

The presence of grimacing or purposeful movements is important. Segmental reflexes alone can cause the limb to move in response to pain.

The body

Look for signs of trauma. Examine the heart, lungs and abdomen.

The urine

Note whether there is incontinence. Test the urine for glucose and ketones (diabetic ketoacidosis), protein (uraemia) and blood (trauma).

The blood glucose

Always prick the finger, place a drop of blood on an impregnated test strip, and test for hypoglycaemia or hyperglycaemia. If this cannot be done immediately, give the patient a bolus of intravenous glucose anyway (which will not usually harm the patient in diabetic ketoacidosis, but will save the life of a patient with hypoglycaemia). If there is any suspicion of Wernicke's encephalopathy, thiamine must be given as well.

The temperature

Hypothermia (e.g. exposure or hypothyroidism) or fever (e.g. meningitis) must be looked for.

Stomach contents

While protecting the airway, examine stomach contents by inserting a nasogastric tube and washing out the stomach if a drug overdose is suspected, or if no other diagnosis is obvious.

suspected, or if no other diagnosis is obvious.

Coma scale

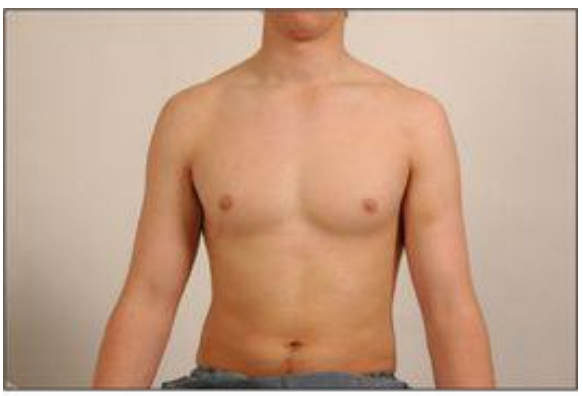
It is most useful to score the depth of coma, as changes in the level of consciousness can then be judged more objectively (Table 11.42).

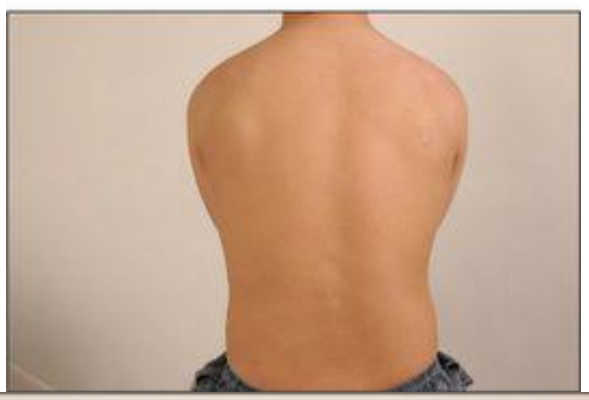
Summary

Examining the nervous system: a suggested method (Figure 11.111)

Handedness, orientation and speech

Ask the patient if he or she is right- or left-handed. As a screening assessment, ask for the patient's name, present location and the date. Next ask the patient to name an object pointed at and have him or her point to a named object in the room, to test for dysphasia. Ask the patient to say 'British Constitution' to test for dysarthria.





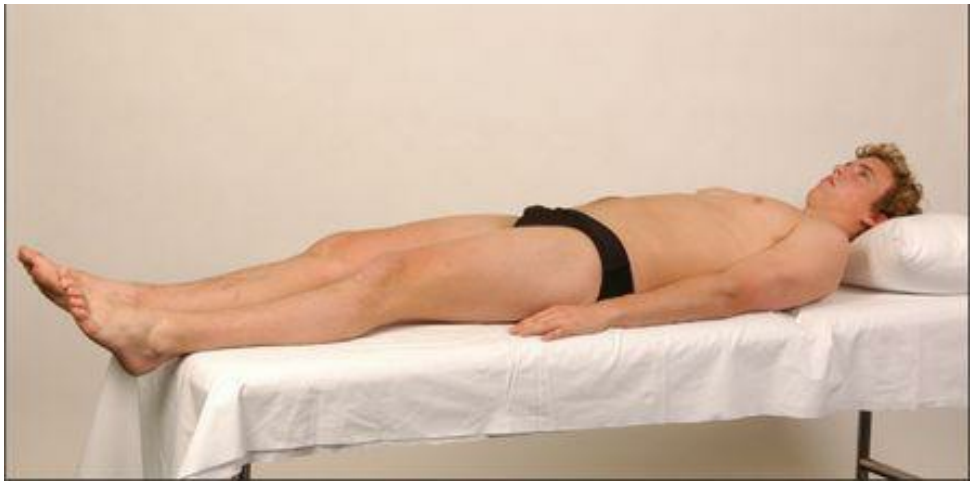


Figure 11.111 The nervous system examination

1. **HIGHER CENTRES EXAMINATION GUIDE**

Lying or sitting

1. **General inspection**

Obvious cranial nerve or limb lesions
Ask patient about handedness, level of education
Shake hands

2. **Orientation**

Time
Place
Person

3. **Speech**

Name objects (nominal dysphasia)

4. **Parietal lobes**

Dominant (Gerstmann's syndrome)

- Acalculia—(mental arithmetic)
- Agraphia (write)
- Left–right disorientation

• Finger agnosia (name fingers)

Non-dominant

- Dressing apraxia

Both

- Sensory inattention

• Visual inattention
• Cortical sensory loss (loss of graphaesthesia, two-point discrimination, joint position sense and stereognosis)
• Constructional apraxia

5. **Memory (temporal lobe)**

Short-term (e.g. names of flowers)

Short-term (e.g. names of flowers)
Long-term

6. **Frontal lobe**
   Reflexes—grasp—pout—palmar mental
   Proverb interpretation
   Smell
   Fundi
   Gait
7. **Other**
   Visual fields
   Bruits
   Blood pressure, etc.

2. **NECK STIFFNESS AND KERNIG'S SIGN**

3. **CRANIAL NERVES**
   - II Visual acuity and fields; fundoscopy
   - III IV VI Pupils and eye movements
   - V Corneal reflexes
   - VII Facial muscles
   - VIII Hearing
   - IX X Palate and gag
   - XI Trapezius and sternomastoids
   - XII Tongue

4. **UPPER LIMBS EXAMINATION GUIDE**
   1. **General inspection** (patient sitting to begin with)
      Scars
      Skin (e.g. neurofibromata, café-au-lait)
      Abnormal movements
   2. **Shake hands**
   3. **Motor system**
      Inspect arms, shoulder girdle—extend both arms
      - Wasting
      - Fasciculation
      - Tremor
      - Drift

      Palpate
      - Muscle bulk
      - Muscle tenderness

      Tone
      - Wrist
      - Elbow

      Power

Power
- Shoulder
- Elbow
- Wrist
- Fingers
- Ulnar, median nerve function

Reflexes
- Biceps
- Triceps
- Supinator
- Finger

Coordination
- Finger–nose test—intention tremor, past-pointing
- Dysdiadochokinesis
- Rebound

4. **Sensory system**
   Pain (pinprick)
   Vibration (128 Hz tuning fork)
   Proprioception—distal interphalangeal joint (each hand)
   Light touch (cottonwool)
5. **Other**
   Thickened nerves (wrist, elbow)
   Axillae
   Neck
   Lower limbs
   Cranial nerves
   Urine analysis, etc.

5. LOWER LIMBS EXAMINATION GUIDE

Patient lying

1. General inspection
   Scars, skin
   Urinary catheter
2. Gait
3. Motor system
   Inspect
   - Wasting
   - Fasciculation
   - Tremor

   Palpate
   - Muscle bulk
   - Muscle tenderness

   Tone

- Knee—and test for clonus
- Ankle—and test for clonus

Power

- Hip
- Knee
- Ankle
- Foot

Reflexes

- Knee
- Ankle
- Plantar

Coordination

- Heel–shin test
- Toe–finger test
- Foot tapping test

4. Sensory system
   - Pain
   - Vibration
   - Proprioception
     - Light touch
5. Saddle region sensation
6. Anal reflex
7. Back
   - Deformity
   - Scars
   - Tenderness
   - Bruits

Neck stiffness and Kernig's sign

Ask the patient to lie flat and attempt gently to flex the head by placing a hand under the occiput. Flex the patient's hip with the knee bent and then attempt to straighten the leg.

Cranial nerves

The patient should sit over the edge of the bed if possible. Begin by general inspection of the head and neck looking for craniotomy scars, neurofibromas, facial asymmetry, ptosis, proptosis, skew deviation of the eyes or inequality of the pupils.

The second nerve

The second nerve

Test visual acuity with the patient wearing his or her spectacles. Each eye is tested separately, while the other is covered with a small card.

Examine the visual fields by confrontation using a hat pin or fingers. The examiner's head should be level with the patient's head. Each eye is tested separately. If visual acuity is very poor, the fields are mapped using the fingers.

Look into the fundi.

The third, fourth and sixth nerves

Look at the pupils, noting the shape, relative sizes and any associated ptosis. Use a pocket torch and shine the light from the side to gauge the reaction of the pupils to light. Assess quickly both the direct and consensual responses. Look for an afferent pupillary defect by moving the torch in an arc from pupil to pupil. Test accommodation by asking the patient to look into the distance and then at the hat pin or finger held about 30 cm from the nose.

Assess eye movements with both eyes first, getting the patient to follow the pin in each direction. Look for failure of movement and for nystagmus. Ask about diplopia in each direction.

The fifth nerve

Test the corneal reflexes gently and ask the patient if the touch of the cottonwool on the cornea can be felt. The sensory component of this reflex is V and the motor component VII.

Test facial sensation in the three divisions: ophthalmic, maxillary and mandibular. Test pain sensation with the pin first and map any area of sensory loss from dull to sharp. Test light touch as well so that sensory dissociation can be detected if present.

Examine the motor division of the fifth nerve by asking the patient to clench the teeth while you feel the masseter muscles. Then get the patient to open the mouth while you attempt to force it closed; this is not possible if the pterygoid muscles are working. A unilateral lesion causes the jaw to deviate towards the weak (affected) side.

Test the jaw jerk. This is increased in cases of pseudobulbar palsy.

The seventh nerve

Test the muscles of facial expression. Ask the patient to look up and wrinkle the forehead. Look for loss of wrinkling and feel the muscle strength by pushing down on each side. This is preserved in upper motor neurone lesions because of bilateral cortical representation of these muscles.

Next ask the patient to shut the eyes tightly and compare the two sides. Tell the patient to grin and compare the nasolabial grooves.

The eighth nerve

Softly whisper a number 60 cm away from each ear. Examine the external auditory canals and the eardrums if this is indicated.

The ninth and tenth nerves

Look at the palate and note any uvular displacement. Ask the patient to say 'Ah' and look for symmetrical movement of the soft palate. With a unilateral lesion the uvula is drawn towards the unaffected (normal) side. Test gently for sensation on the palate (the ninth nerve). Ask the patient to speak to assess hoarseness, and to cough. A bovine cough suggests bilateral recurrent laryngeal nerve lesions.

The twelfth nerve

While examining the mouth, inspect the tongue for wasting and fasciculation. Next ask the patient to protrude the tongue. With a unilateral lesion the tongue deviates towards the weaker (affected) side.

The eleventh nerve

Look for torticollis. Ask the patient to shrug the shoulders and feel the trapezius as you push the shoulders down. Then ask the patient to turn the head against resistance and also feel the bulk of the sternomastoid. Then examine the skull and auscultate for carotid bruits.

Upper limbs

Shake the patient's hand firmly. Ask him or her to sit over the side of the bed facing you, if possible.

Examine the *motor system* systematically every time. Inspect first for wasting (both proximally and distally) and fasciculations. Don't forget to include the shoulder girdle in your inspection.

Ask the patient to hold both hands out (palms up) with the arms extended and close the eyes. Look for drifting of one or both arms (upper motor neurone weakness, cerebellar lesion or posterior column loss). Also note any tremor, or pseudoathetosis due to proprioceptive loss.

- Feel the muscle bulk next, both proximally and distally, and note any muscle tenderness.

- Test tone at the wrists and elbows by passively moving the joints at varying velocities.

- Assess power at the shoulders, elbows, wrists and fingers.

- If indicated, test for an ulnar nerve lesion (Froment's sign) and a median nerve lesion (pen-touching test).

- Examine the reflexes: biceps (C5, C6), triceps (C7, C8) and brachioradialis (C5, C6).

- Assess coordination with finger–nose testing and look for dysdiadochokinesis and rebound.

Motor weakness can be due to an upper motor neurone lesion, a lower motor neurone lesion or a myopathy. If there is evidence of a lower motor neurone lesion, consider anterior horn cell, nerve root or brachial plexus lesions, peripheral nerve lesions or a motor peripheral neuropathy.

Examine the *sensory system* after motor testing, because this can be time-consuming.

First test the spinothalamic pathway (pain and temperature). Demonstrate to the patient the sharpness of a pin on the anterior chest wall or forehead. Then ask him or her to close the eyes and tell you if the sensation is sharp or dull. Start proximally and test each dermatome. Map the abnormal area. As you are assessing, try to fit any sensory loss into dermatomal (cord or nerve root lesion), peripheral nerve, peripheral neuropathy (glove) or hemisensory (cortical or cord) distribution. It is not usually necessary to test temperature.

Next test the posterior column pathway (vibration and proprioception). Use a 128 Hz tuning fork to assess vibration sense. Place the vibrating fork on a distal interphalangeal joint when the patient has the eyes closed and ask whether it can be felt. If so, ask the patient to tell you when the vibration ceases and then, after a short wait, stop the vibrations. If the patient has

deficient sensation, test at the wrist, then elbow, then at the shoulder.

Examine proprioception first with the distal interphalangeal joint of the little finger. When the patient has the eyes open grasp the distal phalanx from the sides and move it up and down to demonstrate, then ask the patient to close the eyes; repeat the manoeuvre. Normally, movement through even a few degrees is detectable, and the patient can tell whether it is up or down. If there is an abnormality, test larger movements and then proceed to test the wrist and elbows similarly if necessary.

Test light touch with cottonwool. Touch the skin lightly (do not stroke) in each dermatome.

Feel for thickened nerves—the ulnar at the elbow, the median at the wrist and the radial at the wrist—and feel the axillae if there is evidence of a proximal lesion. Note any scars, and finally examine the neck if relevant.

Lower limbs

Test the stance and gait first if possible. Then put the patient in bed with the legs entirely exposed. Place a towel over the groin—note whether a urinary catheter is present.

Look for muscle wasting and fasciculations. Note any tremor. Feel the muscle bulk of the quadriceps and run your hand up each shin, feeling for wasting of the anterior tibial muscles.

Test tone at the knees and ankles. Test clonus at this time. Push the lower end of the quadriceps sharply down towards the knee. Sustained rhythmical contractions indicate an upper motor neurone lesion. Also test the ankle by sharply dorsiflexing the foot with the knee bent and the thigh externally rotated.

Assess power next at the hips, knees and ankles.

Elicit the reflexes: knee (L3, L4), ankle (S1, S2) and plantar response (L5, S1, S2).

Test coordination with the heel–shin test, toe–finger test and tapping of the feet.

Examine the sensory system as for the upper limbs: pinprick, then vibration and proprioception, and then light touch. If there is a peripheral sensory loss, attempt to establish a sensory level by moving the pin up the leg and onto the abdomen and, if necessary, onto the chest. Examine sensation in the saddle region and test the anal reflex (S2, S3, S4).

Go to the back. Look for deformity, scars and neurofibromas. Palpate for tenderness over the vertebral bodies and auscultate for bruits. Perform the straight leg-raising test.

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Gates P. The rule of 4 of the brainstem: a simplified method for understanding brainstem anatomy and brainstem vascular syndrome for the non-neurologist. *Intern Med J*. 2005;35:263-266. A simple method is described to localise brainstem vascular syndromes

Khaw PT, Elkinton AR, editors. ABC of eyes, 4th edn. London: BMJ Publishing Group,

2004.

Morris JGL. *The neurological short case*, 2nd edn. London: Edward Arnold; 2005.

Rae-Grant AD. *Weiner and Levitt's Neurology*, 8th edn. Philadelphia: Lippincott, Williams & Wilkins; 2008.

[a] Georges Guillain (1876–1961), Jean Alexandre Barré (1880–1967) and A Strohl described the syndrome in 1916; Strohl's name was dropped because of anti-German feeling during World War I.

[b] Hysteria is an old but still popular term that refers to a presumed psychogenic condition.

[c] Prosper Ménière (1799–1862), director of the Paris Institution for Deaf-Mutes, characterised this condition just before he died of post-influenzal pneumonia. He was an expert on orchids and a friend of Victor Hugo and Honoré de Balzac. He added the grave accent on the second 'e' in his name himself; the acute accent on the first 'e' was added by his son.

[d] James Parkinson (1755–1824), English general practitioner, published an essay on 'The Shaking Palsy' in 1817. He was nearly transported to Australia for reformist activities.

[e] Josef Brudzinski (1874–1917), Polish paediatrician , described this in 1909.

[f] Vladimir Kernig (1840–1917), St Petersburg neurologist, described this in 1882.

[g] The anatomy and function of the cranial nerves was well established by the late 19th century. Galen identified at least seven of the cranial nerves in the 2nd century. These were probably the optic, oculomotor, the sensory part of the trigeminal, the motor part of the trigeminal, facial, vestibular, the glossopharyngeal (including the vagus and accessory) and the hypoglossal.

[h] William Allen Sturge (1850–1919), British physician, described this in 1879, and Frederick Parkes Weber (1863–1962), London physician, described it in 1922.

[i] Samuel von Sommerring (1755–1830) is responsible for the modern classification of 12 cranial nerves. He separated the vestibular from the facial, and the glossopharyngeal from the vagus and accessory.

facial, and the glossopharyngeal from the vagus and accessory.

[j] Other abnormalities of smell are hyperosmia and parosmia. *Hyperosmia* is an increase in the sensitivity of the sense of smell. It is often a sign of psychosis or hysteria but may occur with migraine, during menstruation and in cases of encephalitis. *Parasomia* is a perversion of the sense of smell. It can occur following trauma to the head and in some psychoses. Olfactory hallucinations are more often than not a result of an organic lesion and suggest an irritating lesion in the olfactory cortex.

[k] Hermann Snellen (1834–1908), Dutch ophthalmologist, invented this chart in 1862.

[l] Ludwig Edinger (1855–1918), Frankfurt neurologist, and Carl Friedrich Otto Westphal (1833–90), Berlin neurologist.

[m] Gross differences are abnormal and called *anisocoria*. A small amount of fluctuation in the size of the pupils is normal and called *pupillary unrest*. More pronounced rhythmical contraction and dilatation of the pupils is called *hippus;* this may follow recovery from a third nerve palsy or occur during sleepiness. This is not often of any significance and is not a localising sign.

[n] Robert Marcus Gunn (1850–1909), London ophthalmologist, described the defect in 1883.

[o] William Adie (1886–1935), Australian neurologist working in Britain, described this in 1931.

[p] Henri Parinaud (1844–1905), French ophthalmologist, described this in 1889.

[q] J Steele and J Richardson, Canadian neurologists, described this in 1964.

[r] Johann Laurenz Gasser (1723–65), professor of anatomy, Vienna.

[s] Testing of the sneeze reflex is not routine. Here stimulation or irritation of the nasal mucosa with a hair or small piece of string is followed by contraction of the muscles of the nasopharynx and thorax. The afferent limb of this arc is through the trigeminal nerve and the efferent limb through the facial, glossopharyngeal, vagus and trigeminal nerves, and the motor nerves of the cervical spine. The reflex centre is in the brainstem and upper spinal cord.

[t] Sir Charles Bell (1774–1842), professor of anatomy at London's Royal College of Surgeons, later professor of surgery at Edinburgh, described

facial nerve palsy in 1821.

[u] James Ramsay Hunt (1874–1937), American neurologist.

[v] A patient with an old Bell's palsy may exhibit synkinesis. When the patient blinks the corner of the mouth twitches; when the lips are protruded the affected eye closes.

[w] Alfonso Corti (1822–1888), Italian anatomist, described this in 1851.

[x] Heinrich Adolf Rinné (1819–1968), German ear specialist, described his test in 1855.

[y] Sir Hermann Weber (1832–1918), London physician.

[z] Charles Hallpike (1900–79), English ear, nose and throat surgeon.

[aa] Velum means 'curtain' in Latin.

[bb] Julies Arnold (1835–1915) and Hans Chiari (1851–1916), German pathologists, described this in 1894.

[cc] Aretaeus of Cappadocia reasoned, in 150 AD, that the nerves cross (decussate) between the brain and the periphery and that injury to the right side of the head causes abnormalities of the left side of the body.

[dd] If no fasciculation is seen, tapping over the bulk of the brachioradialis and biceps muscles with the finger or with a tendon hammer, and watching again has been recommended, but this is controversial. Most neurologists do not do this. The reason is that fasciculations are spontaneous. Any muscle movement from a local stimulus is not spontaneous. Even if they occur they may have nothing to do with fasciculations.

[ee] You should not be able to overcome a normal adult patient's power, at least in the legs.

[ff] Sir William Gowers (1845–1915), professor of clinical medicine at University College Hospital, London, and neurologist to the National Hospital for Nervous Diseases, Queen Square, London. He was also an artist who illustrated his own books and had paintings exhibited at the Royal Academy.

[gg] The Queen Square hammer was invented by Miss Wintle, staff nurse at Queen Square. She made hammers from brass wheels covered by a ring pessary and mounted on a bamboo handle; she sold these to medical

students and resident medical officers.

[hh] Actually, dysdiadochokinesis is the inability to perform alternating movements of both wrists with the arms and forefingers extended.*iadochi* is a Greek word meaning succession. The problem here is with successive movements. The Diadochi were the successors of Alexander the Great. They divided his empire.

[ii] Nick Talley has a black belt in Tae Kwon Do and Tang Soo Do.

[jj] In 1826 Sir Charles Bell recognised that there was a 'sixth sense', which was later called proprioception. Vibration sense had been recognised in the 16th century and tests for it developed in the 19th century by Rinné and others. Rydel and Seiffer found that vibration sense and proprioception were carried in the posterior columns of the spinal cord.

[kk] The human dermatomes (which he called pain spots) were first mapped by Sir Henry Head (1861–1940). He was most famous for his experimental cutting of his own radial nerve. This enabled him to chart the order of return of the sensory modalities.

[ll] Albert Ochsner (1858–1925), American surgeon of Swiss extraction, who claimed descent from Andreas Vesalius, the great anatomist.

[mm] Jules Froment (1878–1946), Professor of Medicine, Lyons, France, described the sign in 1915.

[nn] Charles Hoover also described an important sign of chronic obstructive pulmonary disease (page 122).

[oo] Ernst Jendrassik (1858–1921), Budapest physician.

[pp] Josef Babinski (1857–1932), Parisian neurologist of Polish extraction, described this sign in 1896. (It was probably fi rst described by Remak in 1893.) Babinski was a famous gourmet and assistant to Charcot.

[qq] Moritz Heinrich von Romberg (1795–1873), Berlin professor, wrote the fi rst modern neurology textbook. His original description of the sign was of patients with tabes dorsalis (dorsal column disease caused by syphilis).

[rr] Karl Wernicke (1848–1904), professor of neurology at Breslau, described receptive aphasia in 1874. He was killed while riding his bicycle.

[ss] Pierre Broca (1824–1880), professor of surgery at Paris, described this

area in 1861. He described muscular dystrophy before Duchenne.

[tt] Sometimes a word of similar meaning is used (e.g. ‘go’ for ‘start’): this is called *semantic paraphrasia.*

[uu] This syndrome should probably be called ‘*pseudo-pontine-bulbar palsy*’ since the motor nuclei of the fi fth and seventh nerves are in the pons, not the medulla (bulbs).

[vv] Josef Gerstmann (1887–1969), Austrian-born neuropsychiatrist who worked in the United States.

[ww] Sergei Sergeyevich Korsakoff (1853–1900), Russian psychiatrist and great humanitarian, described the syndrome in 1887.

[xx] Euphoria may cause a lack of seriousness, and the repetition of bad jokes and puns (*witzelsucht*)

[yy] Robert Foster Kennedy (1884–1952), a New York neurologist.

[zz] Jacques Jean Lhermitte (1877-01939), French neurologist and neuropsychiatrist.

[aaa] Charles Edouard Brown-Séquard (1817–1894) succeeded Claude Bernard at the College de France. He was the son of an American sea captain (pirate) and a French woman. He was born in Mauritius at a time when it was under British rule. With this background he roved around the world, working in Paris, Mauritius, London and New York. His syndrome usually arose from failed murder attempts. Traditionally Mauritian cane cutters, when trying to murder someone, used a very long thin knife which was slipped between the ribs from behind, to cut the aorta or penetrate the heart. Only such a knife could have caused a cord hemitransection

[bbb] Nicholaus Friedreich (1825–1882), German physician, described this in 1863. He was professor of pathology at Heidelberg. *Pes cavus* is also called Friedreich’s foot.

[ccc] The concepts of hypotonia, rebound and pendular jerks in cerebellar disease stem from Gordon Holmes’ 1917 description of signs in acute unilateral cerebellar disease. They may well not exist in other cerebellar problems. Students will, however, still be expected to know how to test for these signs.

[ddd] Thomas Sydenham (1624–89). He was a captain in Cromwell’s army and became the most famous English physician of his time, providing