Smart handles and red flags in neurological diagnosis

Christopher Hawkes

Eliciting a full history and undertaking a complete neurological examination is time consuming. Knowledge of diagnostic short cuts (smart handles) and caveats (red flags) can speed up this process but care is needed as many tips given here have not been validated scientifically.

The diagnostic process includes pattern recognition, probabilistic, causal or deterministic methods. Simple algorithms, although widely publicized, are rarely implemented by the clinician but in the aeronautical world, for example, algorithms can be used by the inexperienced to diagnose a faulty engine. In this domain, the expert uses 'demons' – diagnostic short cuts recalled from memory – to solve a problem rapidly.

The equivalent to the demon in medicine has been termed 'smart handles' (Hawkes, 1993) from computer parlance 'smart' meaning intelligent and 'handle' referring to a drawer to be opened, hopefully with the answer in it. It is not necessarily the same as pattern recognition, as a smart handle might be a single symptom or sign whereas a pattern is a particular constellation of clinical features. The experienced diagnostician will know many such handles consciously or otherwise. Some have a curious reluctance to divulge their knowledge and would protest it makes a junior doctor's life too easy.

Another diagnostic process is to use caveats or 'red flags' as proposed by Quinn (1995). These are 'negative' handles in the sense that if one comes across a red flag then the diagnosis should be reconsidered. An example is syringomyelia – if the arm reflexes are not absent then the diagnosis is probably wrong. The answer can be reached rapidly by applying a mixture of smart handles and red flags and this approach is presented below. The majority listed here have been passed on from teacher to pupil and very few have been subject to scientific scrutiny. Nevertheless in the observational sense they have proved their worth as a useful way of reaching a diagnosis with speed and often dazzling accuracy. The handles and flags have been grouped into regions based on standard neurological examination and others in relation to specific disease. Any shortcut may be misleading, so the advice should be used with common sense at all times.

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SMART HANDLES IN THE CRANIAL NERVES

Olfactory nerve

Intermittent anosmia: This usually indicates a conductive fault, i.e. air does not reach the olfactory receptor region in the nose because of obstruction by polyps or congestion. Measures that relieve congestion, e.g. ephedrine, steroid sprays, exercise or heavy lifting, temporarily improve the sense of smell. Conversely continuous anosmia is usually perceptive, i.e. a disorder of the olfactory nerve itself or its central connections.

Intermittent olfactory hallucinations: In the absence of mental disorder and local nasal disease this is an ominous symptom, usually indicating a lesion of the anteromedial temporal lobe (uncus) which is a primary olfactory area. The episodes are sometimes called uncinate fits and most often are caused by a malignant glioma.

Normosmia in idiopathic Parkinson's disease: About 80% patients with idiopathic Parkinson's disease (IPD) have reduced sense of smell, especially those with the non-tremulous forms (Hawkes and Shephard, 1998). Not all patients are aware of a smell problem but if on testing (e.g. with a scratch and sniff test) someone with suspected IPD has a normal sense of smell then this is a red flag. You should consider parkinsonism, e.g. progressive supranuclear palsy (PSP), corticobasal degeneration or possibly multiple system atrophy (MSA).

Optic nerve

Bitemporal hemianopia: This is more a localizing sign but it is extremely valuable and can only be found in disorder of the optic chiasm, Apart from a

tendency to collide with doorways the more astute patient may report difficulty in depth perception.

Tunnel vision: There are few organic conditions associated with tunnel vision, e.g. retinitis pigmentosa and temporarily in migraine. The commonest cause is a functional disorder and the patient is usually a young female. Field measurement may reveal a spiral pattern or a constant field size irrespective of screen distance. A genuine field defect is never spiral and enlarges with increasing distance from the screen.

Unilateral dilated pupil in a young woman: This is shown in *Figure 1*. If accommodation is reasonable (although it may be slow, particularly on redilating) and the pupil constricts after prolonged exposure to bright light, it can only be a tonic pupil. If the deep tendon reflexes are depressed then it is called Holmes–Adie syndrome. Many suffer from migraine as well. The pupil often has irregular margins or may be oval shaped. All pupillary reactions tend to be slow and accommodation is usually preserved, but eventually this too is abolished and the pupil remains small and fixed. In the later stages confusion might arise with an Argyll Robertson pupil. The latter is rarely present in isolation, accommodation is brisk and serological tests will put aside any lingering doubt. A completely dilated pupil fixed to all stimuli in an otherwise healthy (often medically qualified) person generally indicates self-administration of atropine, cyclopentolate or similar mydriatic.

Bilateral internuclear ophthalmoplegia: In bilateral internuclear ophthalmoplegia (INO) (*Figure 2*) there is slow movement of the medial recti on conjugate lateral gaze accompanied by nystagmus seen mainly in the lateral rectus muscle ('ataxic' nystagmus). This powerful sign localizes to the medial longitudinal bundle – the dorsally situated pathway which traverses pons and midbrain. It is called internuclear because the lesion lies between the sixth and third cranial nerve nuclei in pons and midbrain respectively. There is a rare pseudo-INO associated with fatiguing ocular muscles in myasthenia gravis, but the vast majority are caused by multiple sclerosis (MS) and a few result from vascular disease particularly if the sign is present unilaterally, i.e. just in one direction of gaze. It has been shown (Athwal et al, 2001) that the pontine centre for bladder control is situated close to the medial longitudinal bundle so that bladder dysfunction often accompanies INO.

Downbeat nystagmus: This consists of a fast downward flick of the eyes followed by a slower upward movement and is best detected on lateral gaze. Normally it indicates a structural lesion at the foramen magnum level (Arnold–Chiari malformation, tumour, syringomyelia) or cerebellar degeneration. Sometimes no obvious cause is found and brainstem ischaemia is presumed.

Third nerve palsy with normal pupillary responses: In a complete third nerve palsy there is ptosis (*Figures 3a, b*), paralysis of all eye movement except abduction (sixth nerve) and intorsion (fourth nerve), with a pupil that is paralysed to all stimuli. Sometimes the palsy is complete except that the pupil reacts normally. This will indicate a medical cause (diabetes, vascular) rather than surgical (aneurysm, tumour). It arises because the pupillary fibres have a blood supply separate from that of the main trunk of the nerve. There are exceptions but it remains a useful guide.

Trigeminal nerve

Jaw supporting sign: This may be obvious only after prolonged conversation and it is performed to support both a weak jaw and neck (*Figure 4*).

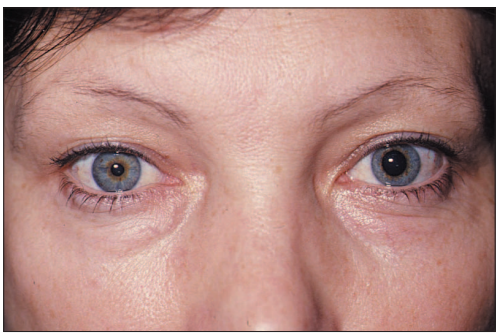


Figure 1. The Holmes–Adie syndrome. The left pupil reacted very slowly and incompletely to light but normally to accommodation, confirming the diagnosis of a tonic pupil.

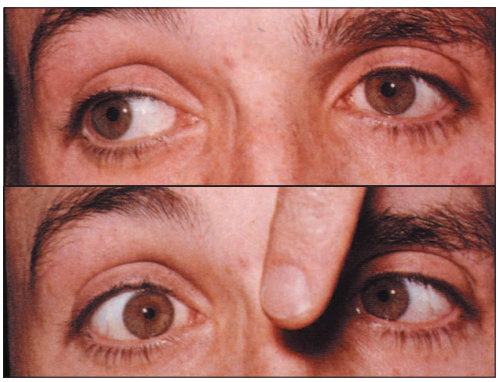


Figure 2. Bilateral internuclear ophthalmoplegia. On looking to either side there is nystagmus of the abducting eye associated with impairment of adduction on conjugate gaze. Adduction may be preserved for accommodation as in this case.

Figure 3. A pupil-sparing ophthalmoplegia of medical rather than surgical origin. Note the normal pupil in the presence of left ptosis. The patient is looking to his right side. The right eye is abducted because of the unopposed action of the intact lateral rectus muscle.

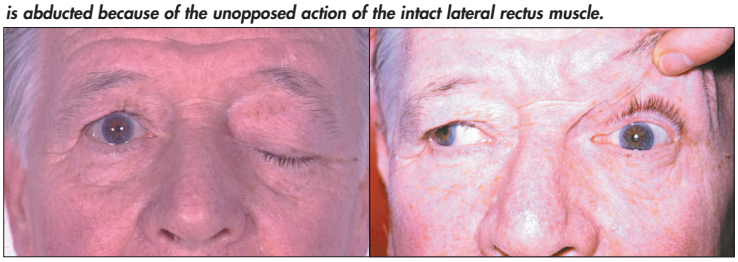


Figure 4. Jaw supporting sign in a patient with myasthenia gravis.

It is pathognomonic of myasthenia gravis where both jaw and neck muscles are weak and fatigue rapidly. The sign is seen also in myotonic dystrophy but there is no fatigue.

Continuous cheek pain: If sinus disease (including malignancy) is excluded then a patient with continuous cheek pain present day and night usually has depression. The typical person is a middle-aged female. Even if depression cannot be substantiated the pain will respond to antidepressants.

Figure 5. Cupid's bow sign. Note the subtle droop of the upper lip on the weak right side.

Intermittent central facial paraesthesia in a young person: This usually indicates hyperventilation. Typically the lips, tongue or nose are involved as well as the extremities. The clue in the history lies in the *periodicity* of the complaint. In most organically based diseases, facial numbness is continuous although its intensity may vary. Care should be taken in elderly hypertensive patients with basilar insufficiency because pontine ischaemia may also produce this. Also metabolic causes have to be considered, e.g. hypoglycaemia and hypocalcaemia. Beware the patient with two conditions, i.e. the anxious patient with intermittent facial tingling superimposed on background numbness.

Geste antagonistique in spasmodic torticollis: Spasmodic torticollis is a form of focal dystonia in which the head turns involuntarily to one side. If the examiner tries to centralize the neck it is usually difficult. Patients often find that gentle pressure from one finger on the chin is sufficient to overcome their head turning. This remarkable phenomenon is poorly understood and not seen in any other disorder. Diagnosis can be made in seconds.

Facial nerve

Cupid's bow sign: This fanciful but memorable sign (*Figure 5*) consists of a droop of the upper lip in patients who have experienced facial palsy of either lower or upper motor neuron variety. Sometimes it is the only evidence of previous facial weakness.

Bilateral facial weakness: If trauma and ear infection are excluded, an acute onset bilateral facial palsy is usually the result of Guillain–Barré syndrome. Longstanding bilateral facial weakness is caused by sarcoidosis, Lyme disease or Melkersson–Rosenthal syndrome. The latter is an autosomal dominant condition characterized by recurrent facial palsy, facial oedema and fissured tongue. Some unlucky patients with idiopathic Bell's palsy have an attack on both sides. If there is bilateral upper facial weakness then leprosy is most likely. The explanation is that the upper face is cooler than the lower and this is thought to provide a better growth environment for the mycobacteria.

Auditory nerve

The patient with a noisy hearing aid usually has bilateral deafness: If the patient has one good ear he/she will turn down the volume of his/her hearing aid. This can be a useful handle if considering an acoustic neuroma; such tumours are unilateral unless there is neurofibromatosis type 2, previously known as central neurofibromatosis.

Accessory nerve and neck muscles

Painless neck flexion weakness: Neck flexion (*Figure 6*) is best examined by asking the patient to flex the neck against the resistance of your hand on his/her forehead. There are four main causes, all beginning with the letter M: myasthenia gravis, myotonic dystrophy, myositis (polymyositis, dermatomyositis) and motor neuron disease. Unfortunately it has become apparent subsequently that the sign is found in other rare myopathies so it is less specific than thought originally. There is a useful red flag: if considering facioscapulohumeral or limb girdle dystrophy and the neck muscles are weak then the diagnosis should be reconsidered as the neck is rarely affected in these conditions. Testing for neck weakness is particularly useful in distinguishing indolent forms of polymyositis from limb-girdle muscular dystrophy.

Stiff neck: a sick patient with negative Kernig's sign: The differential diagnostic list for this is quite short and there is a simple procedure to distinguish the various causes. The most important cause of a stiff neck is meningitis but it also happens with raised intracranial pressure. If there is a Kernig's sign as well then you are probably dealing with meningitis; if this sign is negative then consider raised intracranial pressure. This simple observation can be particularly useful when considering a lumbar puncture – something which would be clearly contraindicated with raised intracranial pressure but desirable in meningitis. Therefore, if confronted with a patient who has neck stiffness but a negative Kernig's sign do a brain scan first before contemplating a lumbar puncture.

Lhermitte's sign: This sign or symptom, provoked by neck flexion, consists of a shock-like feeling which shoots down the spine, occasionally into the arms. It is almost pathognomonic of MS. Rare associations include cervical spondylosis, subacute combined degeneration of the spinal cord, cervical cord tumours and radiation myelitis.

Reversed Lhermitte's sign: A tingling sensation shooting down the spine provoked by neck extension strongly suggests cervical spondylosis. Presumably this is a result of corrugation of the ligamentum flavum (often thickened in spondylosis) which irritates the underlying posterior columns. Rarely patients with MS also report this.

Hypoglossal nerve

Unilateral tongue weakness after car accident: This is rare but if a patient is involved in a road traffic accident typically with whiplash neck injury, and develops unilateral neck pain with tongue weakness after a few hours or at most a few days then carotid artery dissection is the only explanation. The reason is the proximity of the hypoglossal nerve to the origin of the internal carotid artery. Ipsilateral Horner's syndrome sometimes accompanies the dissection.

Florid bilateral tongue fasciculation: If this is a chronic condition, this sign nearly always indicates motor neuron disease of the progressive bulbar variety.

The patient who repeatedly protrudes the tongue: The majority of such people have tardive dyskinesia, i.e. a delayed movement disorder, resulting from exposure to phenothiazine drugs. Characteristically there are associated facial and trunk movements which make this condition quite easy to recognize.

SMART HANDLES IN UPPER LIMBS

Myelopathy hand sign

With the arms held outstretched and supinated, passive abduction of the little finger generally indicates a pyramidal lesion ('myelopathy hand') or ulnar palsy (*Figure 7*). Sensory testing in the hand will resolve which of these is correct: there will be an ulnar pattern of fourth finger-splitting sensory loss in ulnar disease (see below) but a radicular pattern in cervical lesions. Patients with a lesion of the pyramidal tract tend to abduct the little finger passively when the arm is held outstretched. As the defect increases adjacent fingers abduct passively. The sign is often seen in cervical lesions but is not specific for that area and may arise in any pyramidal tract lesion in or above the neck.

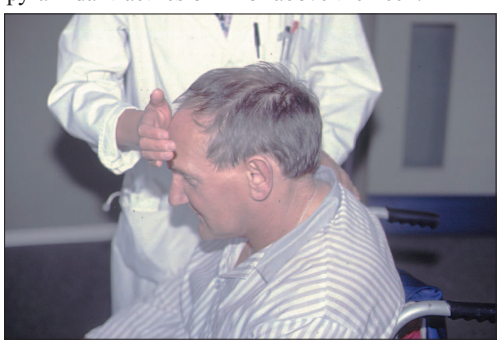


Figure 6. Painless neck flexion weakness. This is a good method of testing the neck muscles. If the movement shown is weak and pain free then only four major causes need be considered.

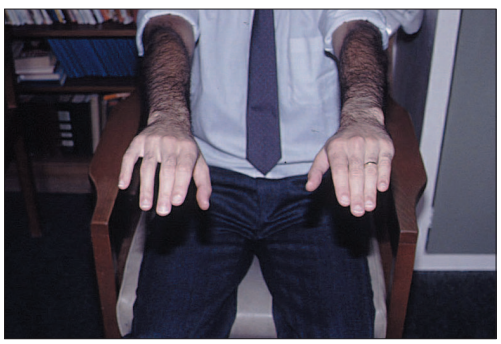


Figure 7. Myelopathy hand sign shown on the right side. There is passive abduction of the fifth finger. This is a sign of pyramidal tract disorder.

Drooped finger sign

If the arms are held outstretched and pronated and one or two fingers are found to droop (i.e. passively flex), this usually indicates motor neuron disease or syringomyelia (*Figure 8*). Brief sensory testing will resolve which it is (there is no sensory loss in motor neuron disease). The sign probably develops because of the patchy nature of anterior horn cell damage. It may occur as a transient phenomenon in neuralgic amyotrophy, in motor neuropathy with conduction block, and in those with tendon rupture as a result of rheumatoid arthritis.

Difficulty flexing the distal phalanx of digits I and II

This can only happen in anterior interosseous nerve palsy (*Figures 9a, b*). This nerve is purely motor and innervates just three muscles, i.e. flexor pollicis longus (flexes distal phalanx of thumb), flexor digitorum indicis (flexes distal phalanx of index finger) and pronator quadratus. Hence the pinching movement required to hold a pen is lost and the short finger flexors are used to oppose thumb and forefinger. There is slight weakness of pronation but sensation is intact. Such patients can squeeze a clothes peg but cannot pick up a pin.

Claw hands, claw feet (pes cavus) and depressed reflexes

This is a favourite short case for exam candidates (*Figures 10a, b*) and most often indicates one type of Charcot–Marie–Tooth disease. If the leg reflexes are increased, syringomyelia is the answer – at least it will satisfy the examiner – but there are rare inherited neuropathies, e.g. Dejerine–Sottas disease or Refsum's disease.

Figure 8. Drooped finger sign in someone with motor neuron disease.

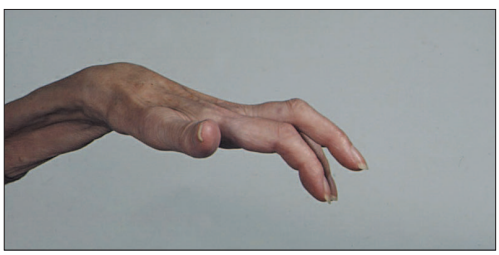
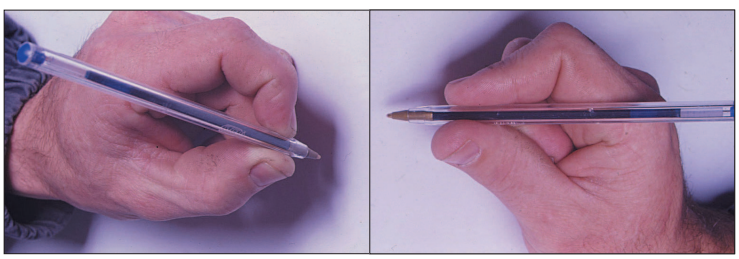


Figure 9. Anterior interosseous palsy. Note the extended distal phalanges of digits 1 and 2 on right side because of weakness of the long flexors (flexor pollicis longus and flexor indicis).



Sensory loss which 'splits' the ring finger

This curious sensory distribution usually indicates an ulnar nerve lesion (*Figure 11*). A lesion of C8 impairs sensation in the little finger and typically the whole of the ring finger. It often extends onto the medial forearm which ulnar nerve lesions never do and may be accompanied by a depressed triceps jerk (C7 and C8).

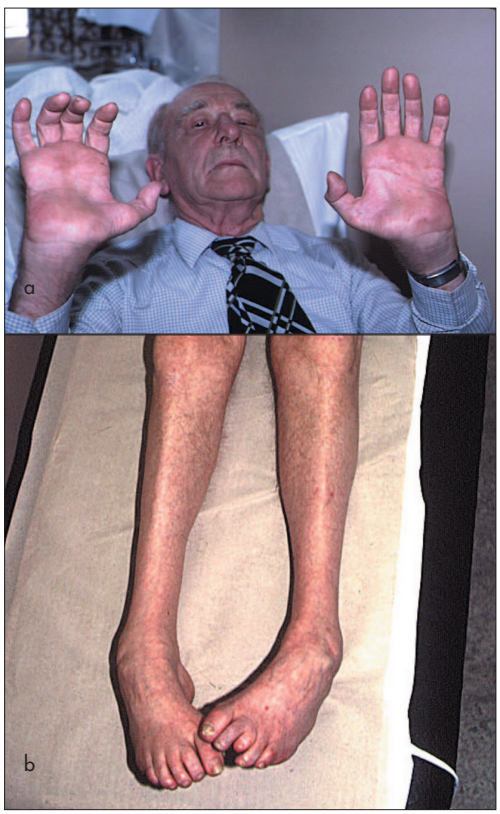
Hand flick in carpal tunnel syndrome

If the patient is woken at night with pain or tingling in the hand and describes a downward flicking movement to relieve discomfort this is strongly suggestive of a carpal tunnel syndrome.

Inverted supinator jerk

In this the normal reflex movement (C6) is replaced by flexion of the fingers (C8, T1). Hence this is a powerful sign localizing the lesion to C6. If the biceps jerk is also inverted, elbow flexion is replaced by finger flexion. Usually the triceps jerk is increased and the cause is most often cervical spondylosis. The likely basis of reflex inversion is simple mechanical spread of the hammer impulse

Figure 10. a. Claw hands. b. Claw feet (pes cavus) and depressed reflexes The patient has Charcot–Marie–Tooth disease (peroneal muscular atrophy or hereditary motor and sensory neuropathy type 1 or 2).



through bone to the nearest functioning reflex. Apart from localizing a lesion it is a useful handle to help distinguish cervical spondylosis from motor neuron disease although it should not be forgotten that the two may coexist.

Rising scapula sign

If the shoulders are both fully abducted, patients with serratus anterior weakness involuntarily elevate the scapula (*Figure 12*), sometimes called the shoulder hump sign. When bilateral this is virtually diagnostic of facioscapulohumeral dystrophy but it can occur in the much rarer scapuloperoneal hereditary neuropathy. In accessory nerve lesions the scapula wings but does not elevate on shoulder abduction because serratus anterior (C5 and C6) keeps the scapula at its correct horizontal level.

Shoulder shrugging sign

In hemiplegic patients without obvious facial weakness it can be hard to know whether the lesion is in the brain or spinal cord. If so, it is often helpful to test repeated shoulder shrugging. If this is normal the lesion will be in the neck. If it is weak or fatigues the damage should be in the brain. This probably happens because the nerves responsible for upper fibres of trapezius are located at C1–C5 and are rarely all involved in cervical lesions.

An easily obtained Babinski response

This usually indicates MS (*Figure 13*). 'Easily obtained' means that it should be elicitable by someone with no medical training. In non-demyelinating lesions of the pyramidal tract the plantar response will also be extensor but this tends to be a rapid movement often associated with withdrawal responses. Some contest the validity of this sign and state that it also occurs in cervical myelopathy. Others consider it to be a sign of longstanding pyramidal disorder so take care.

Striatal toe sign

Here the hallux is extended continuously and often without stimulation although it will appear on walking. It is characteristic of parkinsonian syndromes and must be distinguished from the ordinary Babinski phenomenon.

Exaggerated leg reflexes with normal plantar responses

This apparent paradox seems to occur only in motor neuron disease and the much rarer spinal muscular atrophies. Often the abdominal reflexes are preserved as well. The reason for this is not known but it is a very useful confirmatory handle.

Brisk knee jerks, depressed ankle jerks and extensor plantars

This constellation is well known to exam candidates and usually indicates either subacute combined degeneration of the spinal cord,

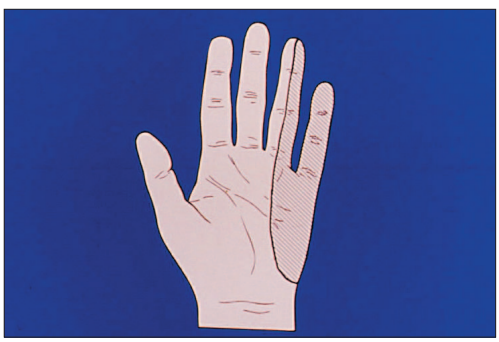


Figure 11. Sensory loss which 'splits' the ring finger. The typical pattern is shown on the palm for ulnar lesions.

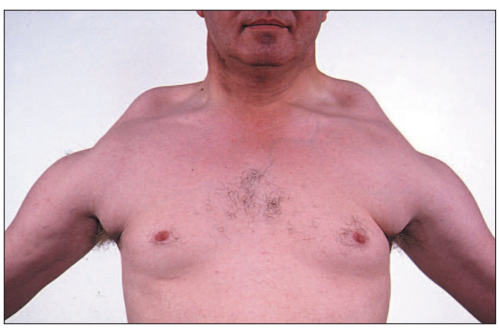
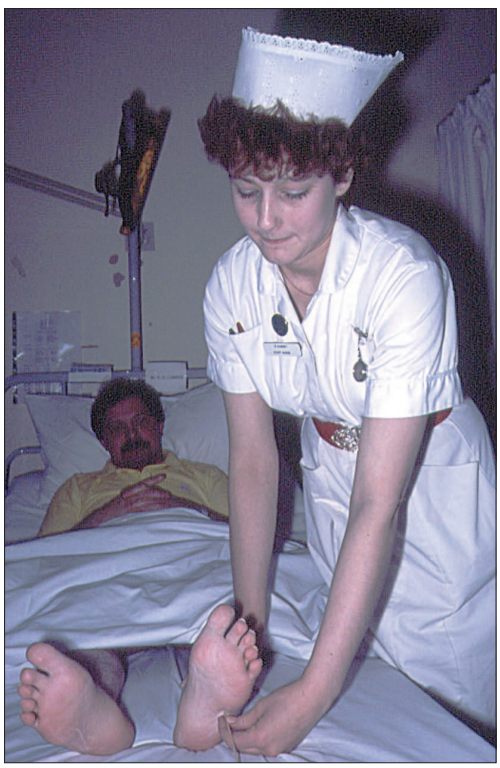


Figure 12. Rising scapula sign. This is shown in a patient with facioscapulohumeral dystrophy.

Figure 13. Easily obtained Babinski response. Babinski sign elicited by a nurse in a patient with multiple sclerosis.



Friedreich's ataxia, motor neuron disease, or taboparesis. All these conditions are associated with a mixture of upper and lower motor neuron pathology. Rarer causes include human immunodeficiency virus-related myeloneuropathy and spinal arteriovenous malformation. Less well known but probably the commonest cause is cervical and lumbar spondylosis which produce a mixture of upper and lower motor neuron signs in the legs.

Bilateral extensor plantar responses with strictly unilateral symptoms

In general this will indicate MS, i.e. the patient complains of a numb or heavy leg and swears the other leg is normal but both plantar responses are found to be extensor. The reason lies in the multiplicity of subclinical plaques in MS, only a few of which give rise to symptoms.

Trembling legs on standing

There is a rare condition known as orthostatic tremor in which patients complain of a vibrating feeling in the legs on standing which lessens or disappears on walking. It is a rapid movement at around 16 Hz. It is difficult to see but may be felt by placing the hands on the thighs while the patient stands. Some have enlarged calves and infrequently there is arm tremor. Many are thought initially to have a non-organic condition and sent to a psychiatrist. This should respond to clonazepam.

HEADACHE HANDLES

Cough headache

This is a headache initiated by coughing or similar manoeuvres that raise intracranial pressure. It must be carefully distinguished from the commonly observed aggravation of pre-existing headache by coughing. About 50% of patients with cough headache have no obvious disease but many of the remainder will have a posterior fossa tumour or cortical venous thrombosis so this is a potentially serious symptom.

Unilateral headache and ipsilateral symptoms

It is worrying to find hemiparesis or hemisensory loss in a patient with headache – vascular anomalies or tumours are obvious considerations. If the symptoms are on the same ('wrong') side as the headache, migraine is the usual explanation.

Continuous headache in a person over 60 years of age

If tension and spondylotic headache is excluded, a continuous headache indicates cranial arteritis until otherwise excluded. The patient usually looks (and may feel) depressed. Paget's disease of the skull also needs consideration.

Morning headache

There are relatively few causes of headache on wakening in the morning that clear later in the day. Tension headache is inclined to start later in the day. If the morning headache is occipital it most often relates to cervical spondylosis or raised intracranial pressure (tumour, systemic hypertension or obstructive sleep apnoea). Inspection of the fundi may reveal papilloedema in the second variety. Morning frontal headache is either caused by sinus disease or migraine. Scalp hypersensitivity often noted while washing the face or combing the hair is a good handle for a migraine headache. Another important cause is chronic carbon monoxide poisoning but the pain is diffuse.

First and worst headache

This is an important red flag. If a patient who is not prone to recurrent headache complains of the first and worst headache in their life, then a serious cause has to be suspected. One would consider vascular tumours, expanding or ruptured aneurysm.

Thunderclap headache

This is defined as headache of abrupt onset as if the head has been hit with a hammer. There are few causes of this of which the most important is a ruptured intracranial aneurysm. This is diagnosed by computed tomography (CT) or magnetic resonance imaging (MRI) brain scan and lumbar puncture. If these tests are negative the headache is likely to be a migraine (including coital headache), cortical vein thrombosis or low-tension headache. Low-tension headache is caused by a dural tear arising spontaneously or after exertion. The cerebrospinal pressure becomes persistently low but it will increase on lying down – as it will in normal people. Its characteristics and mechanism are therefore comparable to post-lumbar puncture headache.

Paroxysmal facial pain

The patient who describes repeated shock-like pain in the cheek or chin which lasts for no more than a second or two is suffering from tic doloreaux. Further questioning usually reveals triggers such as touching the face, eating and cold wind. There is a somewhat similar shock-like pain that affects the forehead or eye and is readily confused with tic doloreaux. The difference is that the shocks last for 15–30 seconds and are associated with conjunctival injection and lacrimation. This is a relatively new syndrome

called sudden unilateral neuralgiform cephalalgia with tearing. It is important to distinguish the two because the response to carbamazepine is good in tic doloreaux but variable in sudden unilateral neuralgiform cephalalgia with tearing.

The patient who wakes repeatedly with severe unilateral orbital pain

Most will have cluster headache. The typical patient is a middle-aged male who wakes around 2 am with excruciating unilateral orbital pain, accompanied by reddening and watering of the eyes and nasal congestion. He has to get out of bed and pace about. The pain recurs at much the same time night after night then disappears spontaneously. Sumatriptan may be effective.

SMART HANDLES ON THE TRUNK

Hemi-sensory loss which includes the trunk

Hemi-sensory loss involving the trunk is likely to mean a deep lesion affecting the thalamus itself. Complete hemisensory loss is also (more frequently) seen in functional disorders and a good discriminatory test is to place a tuning fork on one side of the forehead or sternum and then the other. The patient with non-organic disease will report less vibration on the apparently abnormal side – which is anatomically impossible. Hemisensory loss which *spares* the trunk is frequent in stroke patients and implies a cortical or 'para-thalamic' lesion, probably because of the smaller area of cortical representation for trunk fibres.

Band-like sensations

This is usually reported to be a tight squeezing or tingling sensation around the waist. If it is painful then inflammatory or compressive disease of thoracic nerve roots should be considered. If it is more a gripping feeling or the pain is burning then there are two main causes: MS and vitamin B_{12} deficiency (sub-acute combined degeneration of the cord).

Rapid onset paraplegia

Patients who are admitted as an emergency with rapid onset of paraplegia will have either spinal cord compression typically tumour or prolapsed disc or an inflammatory lesion – transverse myelitis – characteristically caused by MS or a viral infection. As a *general* rule, cord compression or ischaemia will produce paraplegia with few sensory symptoms and signs in the lower limbs whereas an inflammatory lesion will be associated with prominent sensory signs and a good sensory level in addition to the weakness. This happens because sensory fibres are more resistant than motor fibres to compression.

The patient with acute onset of lumbar pain with leg signs suggesting a high lumbar lesion

This is a red flag. Most acute prolapsed discs occur at L5/S1 or L4/L5 and there is weakness of the foot, absent ankle jerks and often bladder disturbance. If the thigh is weak or the knee jerks are absent – suggesting a lesion of L2–L4 – then a disc lesion is unlikely and one should consider other pathology in the cauda equina, e.g. a tumour.

Proximal weakness and dry mouth

If the patient is elderly then Lambert–Eaton myasthenic syndrome should be given serious consideration. The dry mouth is part of a mild autonomic neuropathy, regularly associated with Lambert–Eaton myasthenic syndrome. Some have a tonic pupil as well.

HANDLES FOR MULTIPLE SCLEROSIS

Usually MS is easy to diagnose but there are some more handles that accelerate the process. Lhermitte's sign and easily obtained plantar responses already described above are highly suggestive of MS. Uhthoff's phenomenon is a symptom which consists of worsening of symptoms or signs provoked by raised body temperature or exercise. Patients find they are rapidly fatigued in hot weather but their tolerance is better or normal in cold. Some report deterioration of vision (e.g. enlarging central scotoma or diplopia) after exercise. It is highly characteristic of MS and only very rarely seen in other disease (e.g. demyelinating polyneuropathy). Relapse in the puerperium is also highly characteristic whereas relapse during pregnancy raises a red flag. It would then be appropriate to consider a tumour (e.g. meningioma, glioma, pituitary) or arteriovenous malformation as these are known to enlarge during pregnancy. Therefore if MS is suspected, history taking should include questions relating to these handles.

There are three important red flags for MS:

1. Prominent lower motor neuron signs
2. Conspicuous pain
3. Basal ganglia signs.

Muscle wasting is seen in advanced disease but its presence early on would be most unusual. Central type pain is sometimes complained of in the late stages of disease. This is typically in the lower limbs and has a gripping or burning quality. Spinal pain in MS is most unusual and raises the question of a spinal cord tumour or angioma. If a patient with suspected MS has rigidity or predominant rest tremor that should ring an alarm bell. Cerebellar tremor, i.e. that present on maintaining a posture or undertaking voluntary movement, is frequent in MS but it should disappear with rest.

HANDLES FOR PARKINSON'S DISEASE AND ALLIED SYNDROMES

Idiopathic Parkinson's disease

The cardinal signs are rigidity, akinesia, tremor and postural instability. Lack of upper limb movement may result in repeated stopping of a movement-activated automatic watch, and some patients take their watch to a jeweller thinking it is broken. This is not specific for IPD but is symptomatic of akinesia. By the same token such patients present with a carpal tunnel syndrome affecting the more immobile upper limb.

It is well known that handwriting in parkinsonism becomes small with advancing years (micrographia). Less well known is the tendency to downslope writing. As the disease advances the script slopes toward the right side of the page. Handwriting analysts were able to exploit this fact when presented with the alleged diaries of Adolf Hitler. He suffered from Parkinson's disease in later years and the diaries which were subsequently shown to be forged revealed micrographia but the writing did not progressively slant downwards in successive years. As mentioned earlier, about 80% with IPD have a deficient sense of smell. Anosmia is therefore a useful confirmatory handle. Unfortunately patients rarely complain of a problem with their sense of smell – it has to be tested. If the patient is suspected to have IPD and the sense of smell is normal then the diagnosis should be reviewed – a red flag.

There are other important red flags when considering IPD. If the patient is chair bound within 5 years of onset this would be unusual and it raises the question of a parkinsonian syndrome. Disproportionate antecollis is unusual in IPD. If this is found one should consider MSA or less likely PSP. Finally symmetrical rigidity is not characteristic of IPD, more a feature of the look-alikes such as PSP.

Progressive supranuclear palsy: This atypical form of parkinsonism is characterized by exclusively axial rigidity and akinesia, associated with impaired vertical eye movement and ultimately cognitive decline. Response to levodopa is usually poor. There are three useful handles described by Quinn (1995). When asked to stand up from a chair the patient may do so with almost reckless rapidity and out of proportion to other movement – 'rocket sign'. In the early stages, before vertical gaze is lost, there may be difficulty in moving the eyes straight up. Instead they are moved sideways and upwards in a lateral arc – sometimes called the 'round the houses sign'. Micrographia is seen but can be unusually rapid in contrast to IPD where writing is small and slow. This anomaly has been termed 'fast micrographia'.

Multiple system atrophy: In this disorder there is a mixture of parkinsonism often with early and prominent autonomic dysfunction (Shy–Drager variant). The latter usually consists of bladder problems, impotence and orthostatic hypotension. A striking feature of MSA is cold hands. This is unusual in IPD and probable reflects underlying dysautonomia of peripheral blood vessels. The MRI may reveal a characteristic pontine abnormality the hot-cross bun sign. This results from atrophy of crossing cerebellar fibres which give rise to a cross shape. It has also been termed the X-wing fighter sign depending on your imagination (*Figure 14*).

HANDLES FOR INTERNAL CAROTID INSUFFICIENCY

Atherosclerotic narrowing of the origin of the internal carotid artery is usually diagnosed with ease. Patients present with transient ischaemic attacks and there is often a bruit over the artery's origin. When narrowing is severe (over 95%) then impaired flow develops instead of embolization, and may cause some interesting and powerful handles which apply provided local ocular conditions are absent. The patient may notice monocular blindness (or dimming of vision) on walking outside into bright sunlight. This is presumably caused by retinal ischaemia. By the same token if there is diabetic retinopathy, the retinopathy is less severe on the ischaemic side and the corneal arcus is more prominent in the ischaemic eye. If the internal carotid artery is occluded proximal to the ophthalmic branch then a collateral circulation may develop in the iris via branches of the external carotid artery. This produces reddening of the conjunctive and iris – rubeosis iridis.

Most recently described is orthostatic limb shaking. These patients also have critical narrowing of an internal carotid artery. Blood flow is adequate in the lying or sitting position but on standing perfusion drops and gives rise to shaking episodes in the contralateral arm (rarely the leg) lasting intermittently, sometimes for hours. The shaking can be

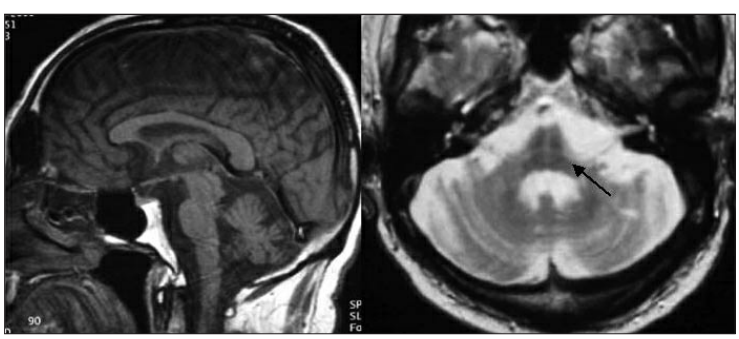


Figure 14. Hot-cross bun sign in multiple system atrophy. Note the white cross-like structure in pons (arrowed).

confused with epilepsy (partial seizures) or parkinsonism. It has to be distinguished from orthostatic tremor (see above) which affects both legs and is rarely visible with the naked eye.

MISCELLANEOUS HANDLES

Distal tingling in polyneuropathy

If a patient complains of tingling in the extremities and a polyneuropathy is being considered then it is probably acquired. In general patients with inherited neuropathy, e.g. Charcot–Marie–Tooth disease, may have distal sensory loss but the patient does not complain of tingling.

Mattress on the floor sign

This occurs when a patient is admitted to a general hospital with epilepsy 'so severe' that the nursing staff feel the patient has to be put on the floor for his/her own safety (*Figure 15*). Pseudo-seizures are the most likely cause. Occasional patients with simulated fits may become violent – often to their medical attendants. Later on they boast how many people were required to 'hold them down' in the attack. Some severe institutionalized genuine epileptics do have to be nursed this way, however.

Blackout with eyes tightly shut

In a genuine blackout the eyes are generally open and staring with pupils dilated and unresponsive. If the eyes happen to be closed there should be no difficulty in opening them. If resistance to eye opening is displayed this will indicate a functional disorder. Furthermore if the patient's eyes are deviated to look at the ground or mattress, it is worth rolling him over to the other side. In a genuine attack the eyes will stay in the same direction; in a simulated episode the eyes will turn to face the floor or mattress again. Some patients with pseudoseizures seem to like wearing large animal-style slippers. I termed this the 'slippers sign' and it should be regarded as a red flag.

Figure 15. Mattress on the floor sign. This occurred in a patient with pseudoseizures.

The pile-free patient who prefers to stand during interview

This is a red flag and usually indicates a person with back disorder of functional origin often pursuing litigation.

The over-friendly patient

This is the patient when seen for the first time remarks something like 'what a nice/wet/cold day it is Doctor'. He/she is unlikely to have organic illness and will often turn out to be neurotic or someone who spends his/her life being referred from one consultant to another.

The patient who cycles for miles but walks only yards

This is an organic symptom which generally indicates lumbar stenosis. The patient presents with a history of claudication on walking but physical examination reveals no vascular insufficiency nor indeed will much be found neurologically. If the patient has discovered that cycling is relatively easy then lumbar stenosis is likely to be present. The reason for this lies in the fact that in the flexed-trunk position of cycling the sagittal diameter of the lumbar canal is increased and nerve compression is lessened. HM

KEY POINTS

- Smart handles are short cuts that suggest one diagnosis or a short differential diagnosis.
- Red flags are warning sighs or symptoms that should encourage you to review your preliminary diagnosis.
- If a mixture of smart handles and red flags is used a diagnosis can be made speedily.
- Very few handles and flags have been scientifically validated so they must be used with caution. In practice they are used by many experienced clinicians and found to be helpful.

I have learnt many handles simply by attending grand rounds and international meetings but I am particularly indebted to many neurologists who have taught me over the years in particular Edwin Bickerstaff, Niall Quinn and the late David Marsden.
Figure 2 *is reproduced from Parsons (1993) by kind permission of Mosby-Year Book Europe Ltd.*
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