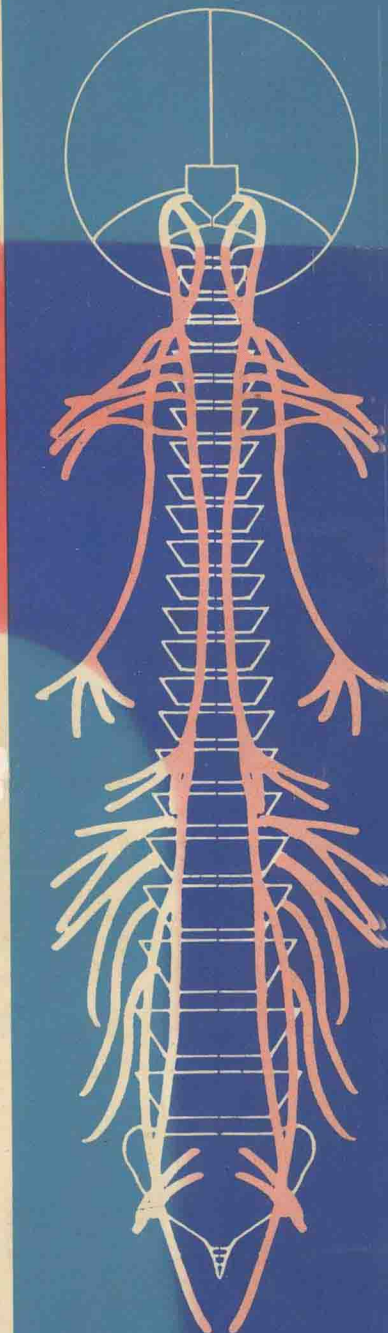
James G McLeod

James W Lance



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Introductory Neurology

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Introductory Neurology

PREFACE

The present book is designed to help the undergraduate medical student relate already acquired knowledge of the basic neurological sciences to the examination of the patient and to diseases of the nervous system. There has been no attempt to discuss in detail all the conditions seen by a neurologist and their treatment since there are many excellent comprehensive textbooks readily available which can and should be consulted. The physiological mechanisms of symptoms and signs are discussed only briefly since the subject has been considered by the authors in another book, *A Physiological Approach to Clinical Neurology* (3rd edn, 1981, Butterworth, London). We are greatly indebted to our colleagues, particularly to Drs R.A. Ouvrier and P.G. Procopis, who have contributed so much by their discussions and in their lectures; to J. Eichorn, L. Hansen and F. Rubiu for the line drawings; and to D. Jones for typing the manuscript.

J.G. McLeod and *J.W. Lance*

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1 HISTORY TAKING

Most patients present to their doctors with a problem, with some observations about themselves or symptoms that they consider abnormal or indicative of ill-health. The first and most important measure in helping patients to solve their problems is to take a detailed unhurried history of the development of symptoms and their evolution, and then to assess this with a knowledge of the patient's family and personal background. In some neurological disorders, it may be possible to reach a firm diagnosis on the history alone. The acquisition of the clinical skills of history taking and physical examination may eliminate the need for further investigations or at least limit these to the minimum necessary to achieve a final diagnosis. The process of history taking involves a number of steps. The precise sequence in which these steps are taken may vary from one medical school to another, from one doctor to another, or even for the same doctor dealing with different problems. The important point is that the thought process should be logical and should lead to a conclusion. Each aspect of the history should be stated as precisely as possible and not glossed over in general terms.

Before the history is taken, the details of the patient's name, address, date of birth, occupation and other necessary data are recorded. After a general enquiry about the purpose of the consultation, notes may be taken about the patient's personal background, family history and past health before going into detail about the problems which chiefly concern him or her. Alternatively, the problem may be analysed carefully as the 'history of the present illness' and then the past, family and personal histories recorded afterwards. The latter approach will be taken to illustrate the case-history of a hypothetical patient, a woman aged 40 years, with a neurological problem.

Presenting symptoms and signs (and their duration)

Weakness of both legs, 2 years; numbness of both legs, 1 year; urgency of micturition, 6 months.

History of the present illness

This section can be presented in note form or as a narrative, recorded as it unfolds from the patient's own account, sharpened a little here and there by an occasional direct question from the clinician to keep the history within the bounds of relevance and to prevent it from degenerating into a list of doctors consulted, investigations ordered and opinions proffered by neighbours and casual acquaintances.

The nature of the symptoms elicited makes the clinician think in terms of localization of the disorder in the nervous system, whether it originates in cerebral hemispheres, brainstem, cerebellum, spinal cord, spinal roots, peripheral nerve or muscle.

The pattern of evolution of symptoms, whether steadily progressive, fluctuating in intensity or remitting for long periods, guides the clinician in assessing the likely pathological cause of the lesion or lesions. In the example given above, the restriction of motor and sensory symptoms to the lower limbs makes one think of a spinal cord lesion and the association with urgency of micturition (disinhibition of bladder reflexes) places the lesion above the conus medullaris, i.e. in the thoracic spinal cord. If the condition is steadily progressive, compression of the spinal cord or a lesion within the spinal cord is likely and warrants immediate investigation. If the condition has fluctuated or remitted one has to entertain the diagnosis of multiple sclerosis. It is important to gauge from the history the *upper limit* of any lesion, if possible. If the patient cited above has also noticed paraesthesiae in the little fingers of both hands, the lesion must be at the level of the eighth cervical segment or above (Fig. 1.1).

Any associated features, precipitating or relieving factors for the symptoms may give a clue as to their nature. This is particularly important in taking a history of any pain, including headache, which should include the following information.

PAIN HISTORY

Length of history: when did the pain first start?
Site: precise part of head, neck, back or other area involved.
Radiation: the distribution of any radicular pain is important for

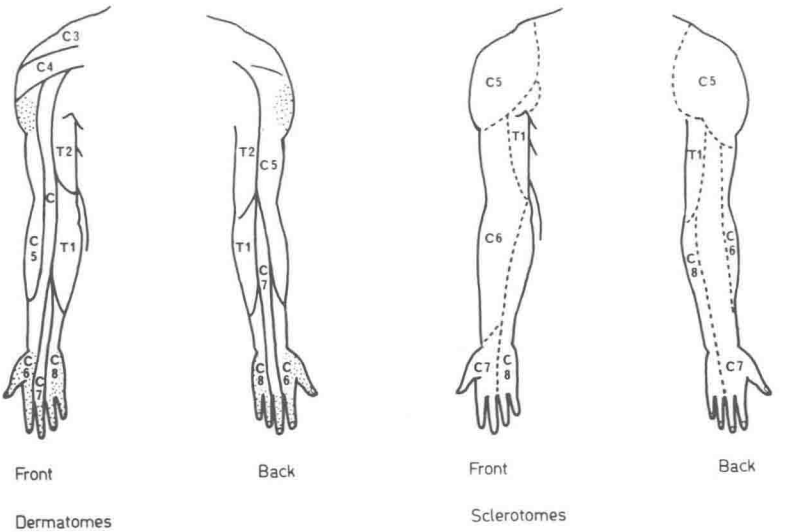

Fig. 1.1 Dermatomes and sclerotomes in the upper limb. If one nerve root only is damaged, sensory loss is usually restricted to the dotted areas shown on the left. Paraesthesiae are generally referred to the dermatome and pain to the sclerotome. (From Lance and McLeod, 1981, by permission of Butterworths, London.)

localization (Figs 1.1 and 1.2), e.g. low back pain may radiate to the medial malleolus (L4 segment), the outer malleolus (L5 segment), the back of the heel (S1 segment). Pain is referred to a sclerotome.

Quality of pain: e.g. stabbing (tic douloureux, lightning pains); throbbing, pulsatile (indicating a vascular component); burning (indicating small sensory fibre involvement); constant, tight, colicky, etc.

Frequency of pain: continuous or intermittent. How many times felt in each day or week?

Duration of pain: seconds, minutes, hours.

Time of onset: any habitual pattern? Awakening from sleep? At the end of the day?

Mode of onset: sudden, slowly progressive or preceded by other symptoms e.g. visual disturbance at the onset of migraine headache.

Associated features: nausea, vomiting, dyspnoea, palpitations or other symptoms.

Precipitating factors: movement of head, neck or back. Coughing, sneezing, straining.

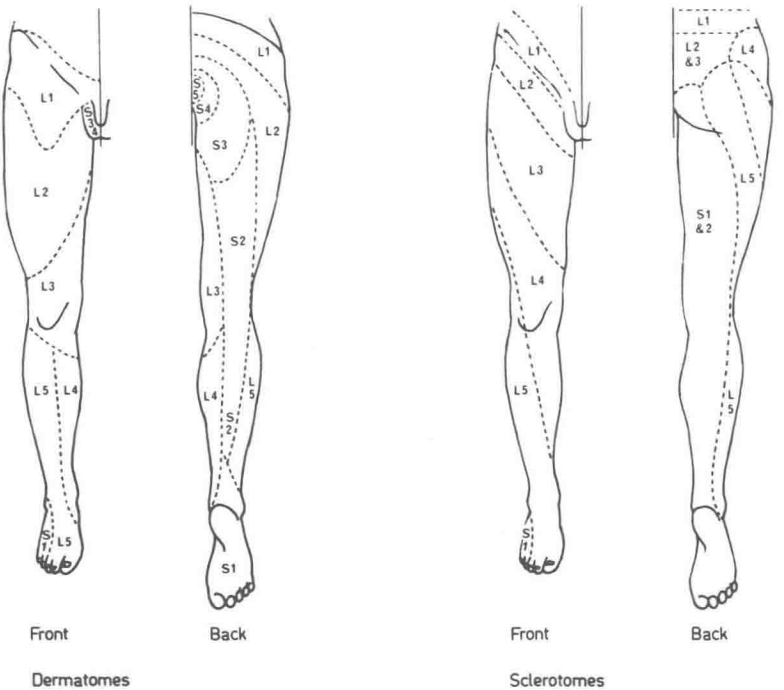

Fig. 1.2 Dermatomes and sclerotomes of the lower limb. (From Lance and McLeod, 1981, by permission of Butterworths, London.)

Relieving factors: certain postures, pressure over the affected part, bed rest, heat or cold, various medications and their effects.

With any syndrome, the symptoms *first* noticed at the onset and the circumstances in which they appeared are of primary importance in determining the site of origin and possible cause.

SPECIFIC INTERROGATION

After the history of the present illness has been clarified as far as possible, the clinician runs through a check list of neurological symptoms to ensure that nothing of relevance has been overlooked, starting at the top of the nervous system and working downwards, asking about the following symptoms:

Intellectual changes, confusion or loss of memory.

Headaches, fits or faints, drowsiness.

Speech disturbance — dysphasia (is the patient right or left handed?); dysarthria; dysphonia.

Sense of smell (cranial nerve 1).

Visual acuity (cranial nerve 2) — impairment or hallucinations of vision; limited to one eye, one half field or bilateral?; details and duration.
Double vision (cranial nerves 3, 4, 6).
Facial paraesthesiae or numbness (cranial nerve 5).
Facial weakness (cranial nerve 7).
If facial palsy is apparent from the history, was it accompanied by hyperacusis or impairment of taste on the affected side?; hearing, tinnitus and vertigo (cranial nerve 8); swallowing, coughing and quality of speech (cranial nerves 9, 10, 12 and central portion of 11).
Weakness — unilateral (hemiparesis: cerebral or brainstem origin), (monoparesis: cerebral, spinal cord or roots); bilateral (quadriparesis: cervical cord or above), (paraparesis; thoracic cord, conus or cauda equina); proximal (myopathy); distal (peripheral neuropathy); fatiguability (myasthenia gravis).
Paraesthesiae or numbness — distribution of same significance as for weakness, with the additional information that (a) numbness of half of the face and opposite side of the body indicates a brainstem lesion; (b) burning sensations down one side of the body with weakness of the other side suggests a unilateral spinal cord lesion (Brown-Séquard syndrome); (c) paraesthesiae from a spinal root disturbance are referred to a *dermatome* (Figs 1.1 and 1.2).
Specific symptoms (common in multiple sclerosis) to be asked about are: (a) tingling down the back on flexing the neck (Lhermitte's sign, electric shock sign) indicating a lower cervical lesion; (b) band-like sensations around the trunk or pelvis; (c) tight wrapping sensations around the legs. The last two symptoms are from dorsal root entry zones or dorsal columns.
Sphincter disturbance — urgency of, or difficulty in initiating micturition or defaecation; incontinence.
Impotence or failure of ejaculation.
Difficulty with coordination, balance or gait.

OTHER SYSTEMS

Since the nervous system does not exist in isolation, the usual questions must be asked about other systems. Those of most relevance to neurological disorders are listed below.

Systemic disturbances, such as connective tissue diseases, malignancy or infections — loss of weight; fever, sweating; aches in muscles or joints.

Respiratory disease (such as carcinoma of the lung, tuberculosis, sarcoid, bronchiectasis) which may be related to neurological symptoms — tightness or pain in the chest; dyspnoea; haemoptysis.

Cardiovascular symptoms, often associated with cerebral vascular disease — palpitations (cardiac dysrhythmias); angina pectoris; intermittent claudication.

Endocrine disorders, drawing attention to the hypothalamus and area around the pituitary fossa — polyuria, polydipsia; symptoms of thyroid or pituitary dysfunction.

Past history

One of the arts of history taking is to know what of the past history is relevant to the present illness or indeed, whether it may actually form part of the present illness. If, for example, the patient whose presenting symptoms and signs were given at the beginning of this chapter eventually disclosed that she had suffered loss of vision in the right eye for a period of 3 weeks at the age of 27 and a tight, band-like sensation around the trunk for 2 months at the age of 35 years, these previous episodes could well be listed at the beginning of the history and would draw attention to the multifocal nature of symptoms experienced by the patient, e.g. transient loss of vision aged 27, band-like sensation around the trunk aged 35, progressive weakness of the legs for the past 2 years.

In the case of epilepsy, mental retardation, or any illness that could have been present at the time of birth or have been initiated by the birth process, the course of pregnancy and labour and birth weight should be noted. Was the baby given immediately to the mother or was resuscitation, intensive care or a humidcrib required? Milestones of development, the time of sitting, standing, walking and talking and progress at school become relevant. Did the child suffer convulsions, with or without a fever? Some 10% of children with febrile convulsions have fits in later life. Head injury, meningitis or encephalitis are obviously of potential relevance to the onset of epilepsy.

A history of headaches or episodes of vomiting ('bilious attacks') in childhood may augur the development of migraine in adult life.

Recurrent headaches with neck stiffness in the past (perhaps diagnosed as meningoencephalitis) may suggest repeated bleeding from a cerebral angioma.

A history of diabetes, hypertension, tuberculosis, immune disorders or other serious illness should be recorded as should any operation, even seemingly minor procedures such as the removal of a 'mole' from the skin.

Family history

Many neurological disorders like Huntington's chorea, the heredo-familial ataxias, Charcot-Marie-Tooth disease, muscular dystrophies and some forms of epilepsy are clearly of genetic origin. Other conditions such as migraine and cerebral vascular diseases tend to 'run in families' although no pattern of inheritance has been determined.

Personal and social background

The personality of the patient, his or her relationship with peer groups, spouse, children, and attitude to occupation may play an important part in the genesis of symptoms considered to be neurological, because of the constant interaction of mind and body. On the other hand, the development of neurological illness may have a profound impact on the patient's ability to work, to maintain satisfactory relationships with family and friends and to derive benefit and enjoyment from life. One must therefore obtain as accurate a picture as possible of the type of life that a patient is leading and hopes to lead in the future to aid in diagnosis and then to help in counselling once the diagnosis is reached. The financial state of the family must also be assessed to see whether some form of financial assistance through sickness benefits or pensions is required. The emotional state of the patient must be carefully assessed as minor degrees of depression are very common and often relevant.

The nature of the patient's occupation and the details of his or her work must be noted because of the possible relevance to the neurological problem and in case of any Workers' Compensation legal action.

Standard questions about the amount of alcohol consumed, smoking habits, exposure to toxins and consumption of drugs are also of importance. The patient should be asked to list all medications currently being consumed or that have been used in the past since they may be relevant to present symptoms and certainly must be known to the clinician before any pharmaceutical agent is prescribed.

GUIDE TO DIAGNOSIS FROM THE HISTORY

The concept of taking such a complete medical history may seem daunting to those who are just starting their clinical training. With greater experience the process becomes easier and quicker, and is spiced with interest or even excitement as the clues present themselves to the receptive mind of the clinician. The phlegmatic recording of the history without any attempt being made by the clinician to correlate the facts as they appear, to make deductions and to form a provisional diagnosis during the process, can be a fruitless exercise. The pieces of the puzzle must be fitted into place while the story is evolving so that any missing fragments can be sought later.

The aim of history taking is to establish as far as possible whether the illness is likely to be of organic or psychological origin. If it is organic, what part or parts of the nervous system may be implicated? What is the pattern of the illness over a period time? What are the precipitating and relieving factors? What is the possible pathology?

Particular attention should be paid to the following points.

Localization

Do the symptoms arise in one site or multiple sites? Can they be explained on the basis of impairment of a particular system (e.g. posterior columns, spinocerebellar pathways, corticospinal tracts) or a particular vascular territory (internal carotid and its branches, vertebrobasilar system, anterior spinal artery)?

Examples of symptoms arising from various sites are:

Cortex Focal (partial) epileptic seizures. Localizing cerebral symptoms (see Chapter 11). Hemianopic or quadrantanopic field defects (see Chapter 8).

Thalamus Contralateral dysaesthesiae (unpleasant burning pain) or sensory loss.

Basal ganglia Parkinson's disease, involuntary movements (see Chapter 10).

Internal capsule Contralateral hemiplegia (and sometimes limb ataxia) and sensory loss.

Midbrain Tectum (superior colliculi): failure of upward deviation of the eyes (Parinaud's syndrome). Tegmentum: third nerve palsy. Contralateral 'wing-beating' (red nucleus) tremor. Contralateral hemiparesis.

Pons and medulla Ipsilateral facial numbness (and often contralateral numbness of the body). Paresis of sixth, seventh, ninth, tenth or twelfth cranial nerves. Dysarthria and dysphagia from involvement of cranial nerves 9–12 is called 'bulbar palsy'. If impairment of speech and swallowing is caused by involvement of corticobulbar pathways rostral to lower cranial nerve nuclei, the condition is called 'pseudobulbar palsy'. Vertigo, tinnitus, deafness (eighth cranial nerve). Ipsilateral Horner's syndrome (sympathetic pathway). Contralateral hemiparesis. Ipsilateral cerebellar signs. Sometimes a lesion in the upper pons may compromise cortico-pontocerebellar fibres so that limb ataxia is on the same side as the hemiparesis ('ataxic hemiparesis').

Cerebellum Midline: ataxia of gait. Hemispheres: ipsilateral incoordination (see Chapter 9).

Spinal cord (see Chapter 7) Bilateral: motor and sensory loss below the level of the lesion. Sparing of posterior columns suggests anterior cord compression or anterior spinal artery involvement. Urgency of micturition. Unilateral: ipsilateral paresis and loss of joint position sense with contralateral loss of pinprick and temperature sensation (Brown-Séquard syndrome). The horizontal level is estimated from any accompanying lower motor neurone lesion, sensory impairment or reflex loss at the segment or segments involved and by the upper limit of the upper motor neuron and sensory tract involvement.

Roots Pain, paraesthesiae, sensory, motor and reflex loss in the distribution of a particular root or roots as described above.

Peripheral nerves (see Chapter 5) Sensory and motor symptoms in the distribution of a particular nerve (mononeuritis), several nerves (mononeuritis multiplex) or all peripheral nerves symmetrically (peripheral neuropathy).

Neuromuscular junction (see Chapter 6) A purely motor syndrome with fatiguability (myasthenia). Muscles commonly involved are:

extraocular muscles (ptosis, diplopia, with *no* pupillary changes); bulbar muscles; proximal limb muscles.

Muscle (see Chapter 6) A purely motor syndrome with proximal wasting and weakness.

Mode of onset

Note the precise sequence of events at the onset of the illness. The initial story given might be one of loss of consciousness. 'I just blacked out, went out like a light'. 'Yes, but what were you doing at the time? Did you notice anything just before you became unconscious?'

Repeating this question in different form a number of times (because patients often think that minor sensations are of no significance and not worth reporting) often helps to diagnose the cause of the episode. Sample replies might include:

'Well, I just turned my head to one side, then my eyes went dim and I felt giddy' (vertebrobasilar insufficiency).

'My heart started racing and I felt faint' (cardiac dysrhythmia causing syncope).

'I just got out of bed to pass urine and woke up on the floor of the bathroom' (micturition syncope).

'My chest was tight and I couldn't get enough air into my lungs' (anxiety hyperventilation).

'I suddenly felt very frightened, as though it had all happened before' (temporal lobe epilepsy).

The principle is to look for the minor symptoms that lead to the major event, for they and they alone may hold the key to the diagnostic doorway.

The evolution (temporal pattern) of the illness

Have the symptoms reached their maximum and then progressively subsided? For example:

Headache — viral meningoencephalitis.

Vertigo, tinnitus — acute labyrinthitis.

Weakness — upper motor neurone: acute stroke; lower motor neurone: Guillain-Barré disease (acute polyneuritis).

Have the symptoms relapsed and remitted? For example:

Headache — migraine.

Vertigo, tinnitus — Ménière's syndrome.

Weakness — upper motor neurone: multiple sclerosis; weakness lower motor neurone: relapsing polyneuritis.

Have the symptoms progressively become worse? For example:
Headache — cerebral tumour or other space-occupying lesion.
Vertigo, tinnitus — acoustic neuroma or brainstem glioma.
Weakness — upper motor neurone and lower motor neurone: motor neurone disease (amyotrophic lateral sclerosis).

The temporal pattern thus gives some idea of the nature of the pathological process.

Conclusion

The history enables an hypothesis to be formed about the site of origin and cause of the patient's symptoms. The physical examination then tests aspects of this hypothesis. After that, further elucidation of the problem may depend on special investigations. Bear in mind that physical examination and diagnostic tests may not help. *Diagnosis often depends entirely on the history.*