THE DIAGNOSIS OF NERVOUS DISEASES II.

By A. D. LEIGH, M.R.C.P.

Registrar, The Maudsley Hospital; One-time First Assistant, Department of Neurology and Psychiatry, The London Hospital

The general principles discussed in the previous article may now be applied to the diagnosis of specific nervous diseases.

I. Motility Disorders

Motility disorders may be considered under four main groups:—

- (a) Spastic disorders of movement.
- (b) Flaccid disorders of movement.
- (c) Transient disorders of movement.
- (d) Involuntary movements.

(a) Spastic motility disorders

Release of the spinal centres and certain brain stem nuclei from cortical control results in the disturbance of muscle tone known as spasticity. Depending on the site of the lesion, one may consider a cerebral group of spastic palsies, and a spinal group.

The *cerebral spastic palsies* may be hemiplegic, affecting one side of the body; diplegic, affecting both sides of the body, especially the lower limbs; and monoplegic, affecting chiefly a single limb. Owing to the large extent of the motor cortex a monoplegia is usually indicative of a cortical or subcortical lesion. In the internal capsule the fibres are so crowded together that a lesion here produces a hemiplegia. If there is a bilateral lesion—most commonly at a cortical or subcortical level, a diplegia results.

Given a cerebral spastic palsy, the differential diagnosis largely depends on the temporal factor—the acuity or chronicity of the process, its congenital or acquired character. Under the eponym of Little's disease are included those cases of hemiplegia or diplegia of pre-natal origin; a history of palsy since birth is diagnostic.

Of the acquired cerebral spastic palsies those of acute onset during adult life are most usually vascular in origin—the result of thrombosis, embolism or haemorrhage. Primary infections of the nervous system, such as acute disseminated encephalomyelitis, or acute disseminated sclerosis are relatively uncommon causes. Neurosyphilis and head injury may be excluded by relevant investigations.

Chronic and progressive hemiplegia is of quite different etiology. A neoplasm, either primary or secondary, is the commonest cause, followed by cerebral abscess, tuberculoma or, extremely rarely, gummatous formation.

The *spinal spastic palsies*, resulting from lesions of the pyramidal tract at a spinal level, form a large group of neurological disorders. Paraplegia of congenital origin is a rarity; in childhood acute paraplegia may result from the disseminated encephalomyelitis of fevers, notably following measles, mumps, and vaccination. A commoner cause however, is Pott's disease, and here the paraplegia may be acute or subacute in onset, the latter being most usual.

In adults disseminated sclerosis holds pride of place, with trauma, syphilis, and acute transverse myelitis of unknown origin entering into the differential diagnosis. A sudden prolapse of an intervertebral disc must be remembered as a cause of acute paraplegia.

Chronic and progressive spinal spastic palsies form the largest group of paraplegias—beginning as a paraplegia in extension and with progression to a complete transverse lesion becoming a paraplegia in flexion. Disseminated sclerosis, neoplasms of cord, roots or vertebrae, motor neurone disease, syringomyelia, syphilis, and the hereditary disorders must be considered, with the emphasis on the exclusion of cord compression.

(b) *Muscular atrophies and flaccid palsies*

In contrast to the preceding group the 'final common path' is the epicentre of the disturbance. Lesions of the anterior horn cell, anterior root, and peripheral nerves may all produce muscular atrophy and flaccid motility disorder. The characteristic objective findings are muscle wasting, motor palsy or paresis, diminished or absent tone and reflexes.

In infancy two conditions of rare occurrence may be encountered, amyotonia congenita and the Werdnig-Hoffman syndrome. The former is characterized by gross loss of power, with flaccidity, with no wasting; the latter by muscular atrophy similar in distribution to the progressive muscular atrophy of adults.

In childhood and adult life the two common causes of an acute flaccid palsy are acute anterior poliomyelitis and, less commonly, an acute toxic polyneuritis. The history, the symmetry of the process, and the spinal fluid findings are diagnostic.

Chronic flaccid motor disorder may result, from disease of anterior horn cell, nerve roots peripheral nerves and muscle.

- (i) Neuronal disorder most commonly occurs with progressive muscular atrophy, medullary neoplasms, and with syringomyelic cavitation. Amyotrophy, fasciculation and fibrillation, and the asymmetry of the signs are of value diagnostically. In both conditions pyramidal disturbance may be observed.
- (ii) Disorders of roots and nerves are to be distinguished by sensory change in dermatome or glove and stocking distribution, by the symmetry of the clinical picture, and by the distal distribution of the muscular weakness. All types of polyneuritis may be encountered, the commonest being diphtheritic in origin. Rarer forms include :—
  - (1) Peroneal muscular atrophy, a heredo-familial disorder, affecting most severely the motor fibres of the nerve trunks, in particular the peroneal nerves. Foot drop, with wasting creeping up the limb to give the 'stork-leg' appearance, followed by wasting of the hand and forearm, occurs characteristically in an adolescent patient.

- (2) Progressive hypertrophic polyneuritis, where the essential diagnostic consideration is gross thickening of the nerve trunks.
- (3) Leprosy—again associated with thickened nerves and perhaps dermal manifestations.

(iii) Disorders of muscle are distinguished by the selective nature of the process, progressive wasting and weakness occurring in certain muscle groups. Beginning in childhood or early adult life, affecting several siblings, there heredo-familial disorders are manifested by a mixture of atrophy and hypertrophy of proximal muscles (hip and shoulder girdles), absence of fasciculation and fibrillation, or of signs of nervous system disease. A rather artificial nosology includes :—

- (1) Pseudo-hypertrophic muscular dystrophy, where the increased bulk of the muscles is the presenting feature.
- (2) Erbs juvenile dystrophy—manifested by simple weakness and wasting of hip and shoulder girdle muscles.
- (3) The Landouzy-Déjerine type of dystrophy—with facial, scapulohumeral and sternomastoid muscles selectively affected.

(c) *Transient or recurrent palsies*

Two uncommon epileptic manifestations, 'Todd's paralysis,' and 'inhibitory' or akinetic attacks, are to be considered in this group. The former follows immediately upon a generalized epileptic attack. Although commonly hemiplegic in distribution, the weakness may affect one limb only. The palsy is usually of only a few hours duration, rarely persisting for a day or two. Full recovery of function always ensues.

'Inhibitory epilepsy' was first described by Gowers and Jackson, transient loss of function being the sole manifestation of the attack.

The cataplectic attacks which occur in association with narcolepsy are an unusual cause of recurrent transient palsy. In addition to the characteristic sleep attacks, sudden accesses of muscular weakness result from

emotional reactions such as laughter, excitement or fear. The patient may fall to the ground, but consciousness is invariably preserved.

In familial periodic paralysis metabolic crises result in recurrent attacks of flaccid paralysis. Heredo-familial in nature and beginning about puberty, the paralysis usually develops during sleep. The patient wakes in the morning to find himself more or less completely paralysed. Consciousness is never impaired. The weakness begins in the hip muscles and spreads upwards, the muscles showing a striking loss of irritability both to mechanical and electrical stimuli. The attack may last for a few hours to several days, and is considered to result from a disorder of potassium metabolism.

Myasthenia gravis is characterized by abnormal fatigability and weakness of the striated muscles, particularly the ocular, facial, pharyngeal and laryngeal musculature. The weakness later becomes generalized. The response to prostigmine has attained diagnostic value, and an injection of 2.5 mgms. of prostigmine should always be used as a therapeutic test. Finally, hysterical weakness or paralysis must be considered. The absence of objective signs of a lesion of the nervous system, and the psychiatric findings are conclusive.

(d) *Involuntary movements*

Six main types of involuntary movements may be recognized, viz., tremors, choreic movements, athetotic movements, torsion spasm, tics, and myoclonic movements.

(i) Tremor

By tremor is meant an involuntary rhythmical oscillation around the point of equilibrium of the affected part. It may be confined to the head, upper or lower limbs, or may be generalized. So diverse are the mechanisms responsible that neurological classification is extremely difficult and for practical purposes a division may be made according to the range of movement into:—

- (1) fine tremors with rapid small oscillations, and
- (2) coarse tremors with slower oscillations.

Into the first group falls the tremor

occurring with thyrotoxicosis, and with intoxications such as mercury, lead, morphine and alcohol. A frontal neoplasm may produce a fine tremor in the upper limbs of the same or of the opposite side.

In the second group may be included the rather coarse tremor of hands and of hepatolenticular degeneration.

Intermediate between the two groups is the rather coarse tremor of hands and face, especially the lips, which is found in G.P.I.

A word might be said here concerning 'intention tremor,' the characteristic feature of which is the accentuation of the 'tremor' by movement. By definition it is not a tremor, but an ataxia—a defect in co-ordination resulting from proprioceptive impairment, whether on the cerebellar or the sensory pathways. The two types of ataxia may be distinguished by the preservation or otherwise of position sense.

(ii) Choreic movements

These sudden, jerky, irregular, involuntary movements of semi-purposive character, involving the face, trunk and extremities, are symptomatic of lesions in the region of the basal ganglia. In childhood they form the presenting feature of Sydenham's chorea; sporadic cases of encephalitic lethargica may also show choreic movements. In adult life Huntington's chorea must be considered, the chronic and progressive course with associated dementia being diagnostic of this condition. Vascular lesions and neoplasms of basal ganglia and superior cerebellar peduncle are encountered more infrequently.

(iii) Athetosis

Athetotic movements are closely allied to choreic movements, and transitional forms are termed choreoathetotic. In contrast, however, the athetotic movements are slow, writhing, irregular and involuntary, being most marked in the upper limb. In early life they are commonly encountered in cases of Little's disease, and in certain rare degenerative

affections. In later life following attacks of encephalitis lethargica, vascular insults and basal neoplasms.

(iv) Torsion spasm

Spasmodic twisting and turning movements of the body, especially of the proximal muscles of hip and shoulder girdles occurs in a variety of extra pyramidal syndromes. It seldom occurs apart from other movements such as chorea or athetosis, and is found in patients suffering from hepatolenticular degeneration, encephalitis lethargica and certain congenital disorders.

(v) The tics

A tic may be defined as a 'sudden, brief irresistible movement, resulting in the involuntary contraction of one or more muscles and representing some act for a particular purpose.'

They may be derived from habits, may be grafted on pre-existing choreic movements, or may be associated with mental conditions such as obsessive compulsive states. Tic-like movements are a well recognized sequelae of encephalitis lethargica.

(vi) Myoclonic movements.

Sudden, local twitching of muscles occurring most commonly in the upper limbs and termed the 'jumps' or 'jerks,' is frequently observed in epilepsy. They may precede the major attacks or be the total representation of an epileptic attack.

II. Disorders of Sensations

Sensory disturbance may be either subjective, the patient complaining of pain or paraesthesiae, or objective, when the examiner may discover areas of analgesia or hypalgesia (loss or diminution of pain sensibility), anaesthesia or hypaesthesia (with reference to light touch). There may be excessive reaction to the different stimuli—a hyperalgesia or hyperaesthesia.

Whatever the variety of sensory disturbance, however, its significance can only be assessed by the consideration of:—

(a) The nature of the sensory change.

(b) The distribution of the sensory change.

(c) The degree of sensory change.

The site of the lesion may then be predicted with considerable accuracy.

(a) *Lesions of peripheral nerves*

A knowledge of the cutaneous areas supplied by individual nerves is essential for accurate diagnosis. In general, however, a small area of analgesia surrounded by a larger area of anaesthesia is suggestive of a peripheral nerve lesion. Trauma or pressure are the commonest causes; leprosy is rarely encountered in this country.

When there is a generalized affection of peripheral nerves—a polyneuritis—the sensory changes occur symmetrically over the distal parts of the limbs. The common causes of polyneuritis have been discussed in the section on flaccid palsies.

(b) *Lesions of spinal nerve roots*

In contrast to a peripheral nerve lesion, with a root lesion the area of analgesia is larger than the area of anaesthesia. This is a result of the overlap of tactile fibres, so that even in extensive lesions of the roots no change in tactile sensibility may be detected. The distribution of the analgesia is in the dermatome supplied by the root concerned, hence the importance of a working knowledge of the dermatome areas.

The common root lesions are produced by tabes, herpes zoster, trauma, or by the effects of pressure whether from a neoplasm of cord, root or vertebral structure. In recent years a prolapsed intervertebral disc has been recognized as one of the commonest causes of root compression. A syphilitic radiculitis, as distinct from tabes, may produce root signs.

(c) *Lesions of the spinal cord*

Loss or impairment of all forms of sensation below a given dermatome is characteristic of a spinal cord affection. The level of the lesion can most easily be obtained by testing with the drag pin, passing from the area of sensory change to the area of normal sensation. With a central intramedullary lesion, pain and thermal sensibility are more severely affected than tactile appreciation.

The common causes of sensory change at the spinal level are :—

- (i) Any cause of pressure upon the cord—whether due to trauma, neoplasm or infective process.
- (ii) Intramedullary lesions such as syringomyelia, neoplasms or disseminated sclerosis.
- (iii) Infections of the nervous system producing a myelitis.

(d) *Lesions of the brain*

A sensory change over one half of the body indicates a lesion on the opposite side of the brain. Whether cortical, subcortical, thalamic or brain stem in situation may be determined by the nature of the sensory change and the presence of other neurological signs. With brain stem lesions crossed paralysis accompanies the sensory change. Thus a lesion in the pons is manifested by a sixth nerve palsy on the side of the lesion and pyramidal and sensory signs on the opposite side.

A profound loss of both cutaneous and deep sensibility is found with a lesion in the neighbourhood of the thalamus. The thalamic syndrome first described by Déjerine and Roussy includes, in addition to the above sensory change, spontaneous pains and an over reaction to certain stimuli such as pain and temperature, accompanied by choreoathetoid movements, especially in the contralateral upper limb.

Loss or impairment of the discriminative aspects of sensation indicates a cortical situation. The maximal sensory change is found over the hands and feet, and there is often little change over the trunk. When a change is observed it can be noted to extend a centimetre or so across the mid line. With a cortical lesion there is a great variability in the responses, as the sensory cortex tires easily.

The usual cause of these sensory syndromes is a vascular insult—commonly a thrombosis or haemorrhage. Neoplasms, primary and secondary, and the demyelinating diseases (disseminated sclerosis, Schilder's disease, etc.) must be excluded.

III. Attack Disorders

Sudden, reversible, recurrent disorder of nervous function occurs in a large number of

conditions of both nervous and systemic origin. The history is the most important diagnostic agent, as in most cases physical signs are equivocal. The age of onset, the character of the attack, and its periodicity are the most useful criteria.

The immature nervous system reacts more violently to the impact of disease than does the mature organization. Hence convulsions in infancy may have a completely different significance to convulsions occurring in later life. In the first few days of life, injury or congenital defect of the brain are the commonest cause of convulsions. Between six months and three years tetany, often precipitated by an attack of diarrhoea or a mild infection, is a potent cause. A convulsion may usher in an exanthem such as measles or a meningeal infection. When repeated convulsions, for which no cause can be discovered, occur in a child, epilepsy must be envisaged as a diagnosis.

During adolescence epilepsy usurps the place of tetany and acute infections. Focal attacks with all that they imply, hypoglycaemic crises, and renal inefficiency begin to assume a role in differential diagnosis.

In adult life episodic attacks of neural dysfunction usually indicates an epileptic disorder. The epilepsies remain one of the most fascinating subjects in neurology, a mirror of the most diverse nervous function. In spite of recent technical advances such as the electro-encephalogram, there is no substitute for wide clinical experience.

A recapitulation of the diverse clinical manifestations of epilepsy is not indicated in a paper of this nature. Suffice it to mention certain important features :—

- (i) An epileptic attack may be considered, in the words of Hughlings Jackson, as due to 'occasional, sudden, excessive discharges of grey matter.' Depending on the degree of spread of this discharge, so an attack may remain focal (Jacksonian) or may become generalized.
- (ii) This explosive discharge may occur at almost any level in the nervous system—but is commonest at the cortical level.
- (iii) Epilepsy cannot be considered as a disease *sui generis*, for it may be the expression of many different conditions—

renal insufficiency, vascular disease, etc.

- (iv) A differentiation into essential or idiopathic and symptomatic epilepsy is useful, although the former group is contracting with advances in knowledge.
- (v) It is useful to consider an epileptic attack in relation to four sequences in times. First, the prodromal period, with its signals of an oncoming fit occurring several hours or days before the attack. Secondly, the aura, which is in effect a part of the attack and immediately precedes loss of consciousness. It is an indication of the situation of the disturbance. Thirdly, the content of the fit, whatever it may be, and fourthly, the sequelae, sleep headaches, limb weakness or automatisms.
- (vi) Disease of the brain, in the sense that pathological changes can be detected post-mortem, may produce either focal or generalized convulsions. In 50 per cent. of patients with a frontal lobe neoplasm, for example, fits occur, and may be quite indistinguishable from the idiopathic variety. It is a sound rule that when an attack occurs in a patient over the age of 40, disease of the brain must be thoroughly eliminated before diagnosing idiopathic epilepsy.
- (vii) Distinction from other paroxysmal disorders.
  - (1) Faints, or syncopal attacks may be extremely difficult to differentiate from petit mal attacks. However, the relation of syncope to emotion, pain or fatigue, the sweating and nausea which precede a faint, and the absence of incontinence are helpful. In Stokes-Adams's attacks the presence of heart block is diagnostic.
  - (2) Menières Syndrome. Here recurrent attacks of violent vertigo, accompanied by vomiting, in a person over 40, are usually diagnostic. The tinnitus and nerve deafness, the long duration of the attack with no impairment of consciousness, are unmistakable. It must be pointed out, however, that vertigo may be an epileptic manifestation, but it is then

usually accompanied by some clouding of consciousness.

- (3) Hysteria may manifest itself by 'attacks'—the hysterical fit so graphically described by Kinnier Wilson 'with limbs quivering and breast heaving, tremulous eyelids and flushed face, the hysteric gives vent to cries, or talks and mutters, resisting any effort to compose her and often needing restraint.' There is no incontinence, the corneal reflex is retained as a rule, and the plantar response is invariably flexor.
- (4) Of the remaining attack disorders, migraine alone requires mention. Here periodic attacks of headache occur, usually unilateral, preceded by visual auras (teichopsia, scintillating scotomata) and accompanied or ending in nausea and vomiting. Hemi-anopic manifestations are well recognized as a migrainous phenomenon but call for intensive investigation to exclude structural changes, in particular angiomatous malformations.

Disorders of Consciousness

Awareness of the environment is a function of the whole organism, in particular of the nervous system working as a whole. It is abundantly clear, however, that certain formations of the brain are more concerned than others with this state of awareness or 'consciousness.' The region surrounding the third ventricle, in particular the hypothalamus, and the brain stem, especially the mid-brain, are important loci for the maintenance of consciousness. Thus diseases affecting the whole brain or, more strictly, focal processes may both produce impairment of consciousness, ranging from mild degrees of confusion to a state of absolute unconsciousness or coma.

Neurologically, however, we may consider four main groups of conditions which cause impairment of consciousness.

(a) Traumatic

The possibility of head injury should be considered in every unconscious patient, for

adequate surgery may be life saving. Concussion and cerebral contusion account for the majority of cases and call for no special discussion here. Extradural haemorrhage is an urgent indication for operation, and here it is well to point out that the 'lucid interval' may vary tremendously from a few minutes to several days. It may be altogether absent. A rapidly deepening coma, with spreading signs, following a head injury awakens suspicion. The diagnosis should be made before the classical Hutchinsonian pupil has appeared.

A subdural haematoma may be acute or chronic in onset, the latter type concerning us here. Headache, drowsiness and mental changes with a history of a fairly recent head injury form the elements of the syndrome. An important feature is the remarkable fluctuation in the signs and symptoms. Diagnosis, however, is not always easy, a history of head injury may not be forthcoming and the semeiology is widely variable.

Lastly, confusion or coma may result from fatty embolization of the brain following trauma to soft parts or bones. The somatic injury, skin petechiae, clinical and radiological signs of 'bronchopneumonia' and the presence of fat globules in the sputum and urine are diagnostic.

(b) *Toxic and metabolic causes*

Alcohol, the barbiturates, the bromides and carbon monoxide are the commonest toxic substances encountered in practice. The effects of alcohol range from the coma of acute intoxication to the confusion found in Korsakow's psychosis. It is important to err on the side of caution before indicting alcohol as the sole causal agent of a coma. Carbon monoxide and the barbiturates are commonly employed in suicidal attempts—the circumstances are often diagnostic.

Bromidism calls for further remark. The widespread and often indiscriminate use of the bromides is to be reckoned with when confronted by an obscure case of mental confusion. A blood bromide estimation (normal .5-2.5 mgms. per cent.) may be revealing. Values below 100 mgms. per cent. may be ignored, but with levels between 100-200 mgms. per cent. symptoms are likely to occur in elderly patients, or in those with renal or cardiovascular impairment.

Of the metabolic disorders diabetes mellitus, hypoglycaemic attacks (both spontaneous, and following the use of Insulin) renal impairment, and liver damage may all produce impairment of consciousness. Two vitamin deficiency states are of importance—Wernicke's encephalopathy resulting from a B.1 deficiency, and pellagra, a deficiency of several factors of the B group. Wernicke's encephalopathy is of interest owing to the focal nature of the lesions, which are found in the hypothalamus and upper part of the mid-brain. Drowsiness and confusion are associated with nuclear oculomotor signs, and may occur in any condition producing a vitamin B.1 deficiency—chronic alcoholism, gastric neoplasm, or starvation.

(c) *Vascular disorders*

Cerebral haemorrhage, thrombosis and embolism are banal causes of impairment of consciousness. The speed of onset and the progression are important differential considerations.

Subarachnoid haemorrhage results in a clear cut syndrome of sudden onset, severe headache, neck rigidity, confusion or coma, and a bloody spinal fluid. Finally thrombophlebitis of the cerebral venous system, spreading from infected ear, nose or sinus commonly goes unrecognized.

(d) *Diseases of the nervous system*

Neoplasm, abscess, acute infections of brain or meninges commonly impair consciousness. Epilepsy and narcolepsy have been discussed previously. It remains to mention some psychiatric conditions—catatonic stupor, the so-called toxic confusional states, and hysterical stupor.

In conclusion, when confronted with the unconscious patient, attention should be directed to the following:—

- (i) The colour of the patient, his breathing, and the smell of his breath.
- (ii) A careful search for any sign of injury.
- (iii) Always test the urine, take the blood pressure and examine the fundi.
- (iv) Careful exclusion of organic disease before diagnosing acute alcoholism or epilepsy.