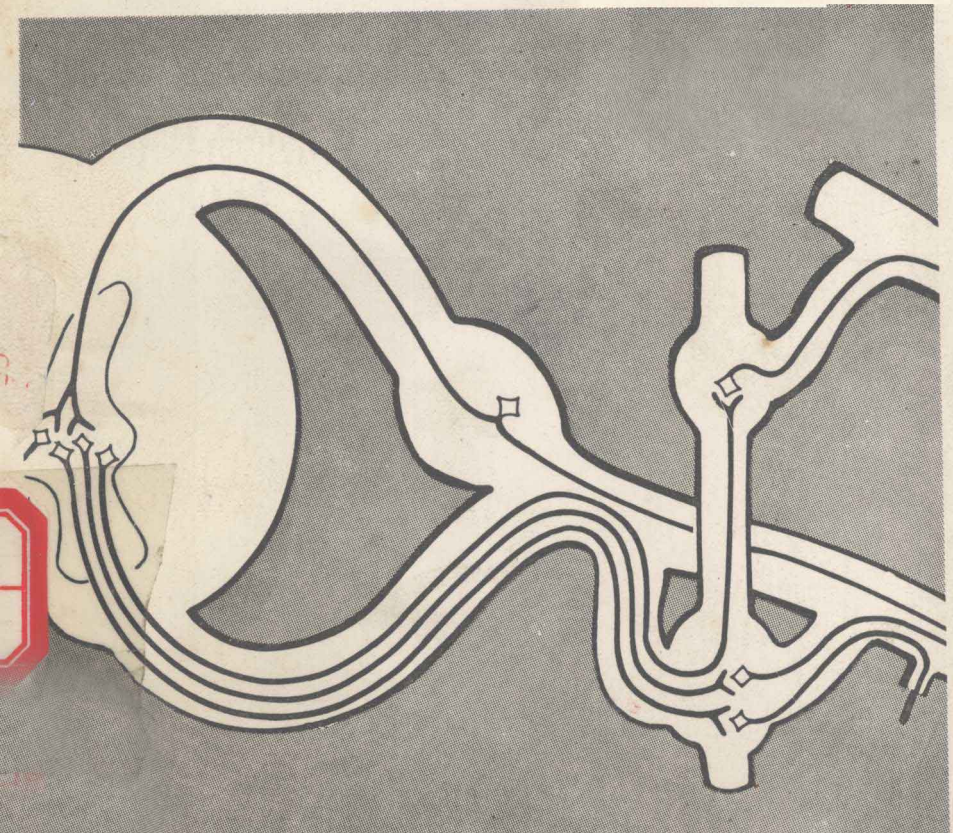
Ivan T. Draper

Lecture Notes on Neurology

Fifth edition



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Preface

Clinical neurology is founded upon the analysis of an accurate history and examination, interpreted in the light of a knowledge of anatomy and of the common neurological illnesses. These notes are intended to provide a basis for this exercise. They were designed for use in conjunction with a formal course of instruction and as a means of rapid revision. This format imposes limitations on the amount of detail which can be provided and it makes no allowance for controversial opinions. My approach to neurology owes a great deal to my own teachers, especially to Professor J.A. Simpson and the late Dr J.B. Stanton, and to my colleagues at the Institute and elsewhere.

I.T.D.

Contents

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LECTURE NOTES ON

Neurology

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Part I
The Structure and Function of the Nervous System

Chapter 1
The Motor System: Organization and Function

The complexity of human behaviour, the range of man's imagination, the accuracy of his perception, his speech and the precision and power of his movements are products of a normally functioning nervous system. The basic unit of the nervous system is the nerve cell or neuron of which there are 10–50 000 000 000. The neuron is a relatively simple structure whose response is limited to the generation of a standard impulse. The variety and adaptability of neural function is achieved by the almost limitless number of connections which exist between the neurons. Each neuron has several fibrous projections, one of which, the axon transmits the impulse generated in the nerve cell body. There are several branching dendrites from which stubby spines project. These form the receptor surfaces for the synapses between one neuron and the next. The spines can accumulate energy from a number of subthreshold stimuli from one axon or from different axons. Eventually the spine discharges and an impulse is transmitted to the cell body. This mechanism of cumulative responses adds spatial and temporal dimensions to neuronal activity. Chains and networks of neurons are activated for various receptor (sensory) and effector (motor) functions.

The execution of precise rapid movement depends on the integrated activity of the whole nervous system, both sensory and motor. It is the custom to divide the motor system into pyramidal and extrapyramidal parts, but it is wrong to equate this division with voluntary and involuntary mechanisms. Of the pyramidal tract fibres, only a quarter originate in the motor cortex, while the rest come from the parietal lobe and the basal–reticular complex. On the other hand, the extrapyramidal system is responsible for a great proportion of voluntary and semi-voluntary movement, particularly the learned and habitual movements, and those associated with posture. Thus the fibres arising from the giant cells of the *motor cortex* are responsible for consciously willed activity, such as the control of fine, unfamiliar hand movements. For such

action to be effective it must be superimposed on the pre-existing postural patterns established by the activity of the basal–reticular system.

The somatic structures are represented over the cortical motor strip, which lies anterior to the central sulcus of the brain (Fig. 1). Electrical stimulation of this area causes movement in the equivalent contralateral part of the body.

The corticospinal tract, comprising fibres from the motor cortex and the parietal lobe, passes down through the internal capsule to the brain-stem. Here it is joined by fibres from the basal ganglia and reticular formation. In the medulla they are grouped into well-defined bundles called the pyramids, and here the major decussation (crossing) occurs. In the spinal cord the crossed pyramidal tracts lie lateral to the central grey matter. The individual fibres finally synapse with the anterior horn cells of the cord, either directly or through one or more interneurons.

The *extrapyramidal system* is less clearly defined. Fibres from the motor cortex synapse with the cells of the basal ganglia, thalamus and reticular formation. Apart from the contribution of

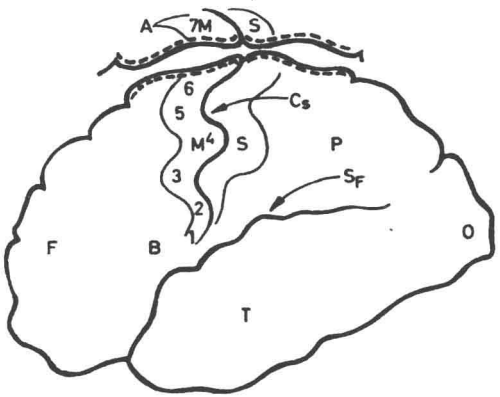

Fig. 1. Lateral view of the left cerebral hemisphere, with the adjacent part of the medial surface shown inset.

A. Accessory motor area. B. Broca's area. Cs. Central sulcus. F. Frontal lobe. M. Motor cortex. O. Occipital lobe. P. Parietal lobe. S. Sensory cortex. Sf. Sylvian fissure. T. Temporal lobe.

Representation of the body over the motor cortex.

1. Mouth. 2. Face. 3. Thumb. 4. Hand. 5. Arm. 6. Trunk. 7. Leg.

fibres to the pyramidal tracts, the majority travel to the cord by the reticulospinal and rubrospinal pathways. The final synapse is with the anterior horn cells, particularly those of the gamma system. Due to the large number of synapses the extrapyramidal system conducts impulses slowly, but by the same token, complex patterns of nerve and muscle activity can be initiated.

Functionally, it is convenient to think of the cerebral cortex influencing and modifying the action of the basal ganglia, which in turn modify the centres of the brain-stem, in turn modifying the cord reflexes. At every level there is a feedback of 'information' to the higher centres (Fig. 2). The cerebellum is a monitor in this feedback network. Disease, at any level in the motor system, releases the centres below from the influence of those above. In determining the level or site of a lesion, the concepts of an upper motor neuron and a lower motor neuron are used.

The upper motor neuron should be thought of as all the descending fibres, both corticospinal and reticulospinal, which

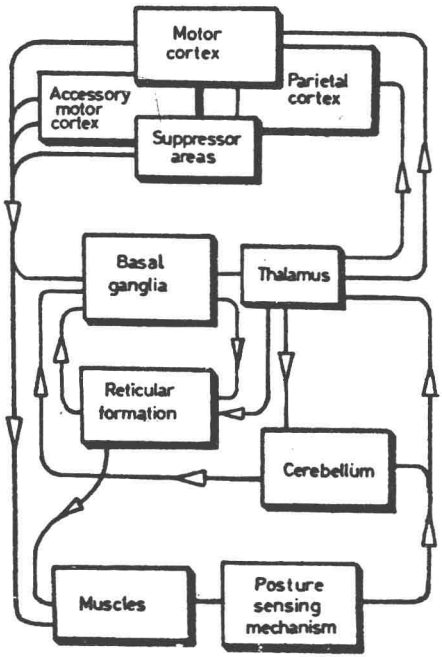

Fig. 2. Block diagram of the motor control system with feedback pathways.

influence the anterior horn cells. The lower motor neuron is the final common path in the motor system, the anterior horn cell of the cord and its axon extending out in the peripheral nerve.

The normal function of the *peripheral nerve* is dependent upon the 'health' of its parent cell body and the integrity of the axon and its myelin sheath. Disordered metabolism in the cell is thus manifest by functional failure at the periphery. Nerve conduction studies provide a method for the in-vivo study of motor nerve function. Each anterior horn cell innervates several muscle fibres. The cell, the axon, its branches and the muscle fibres are termed a motor unit.

The *neuromuscular junction* is bridged by the release of acetylcholine. The arrival of a nerve impulse causes the discharge of minute quantities of acetylcholine from vesicles near the nerve ending. These combine briefly with the receptor protein of the motor end plate—a specialized part of the muscle membrane.

At rest, the inside of the muscle cell carries a negative charge (—90 mV) with respect to the surrounding tissue fluid (Fig. 3a).

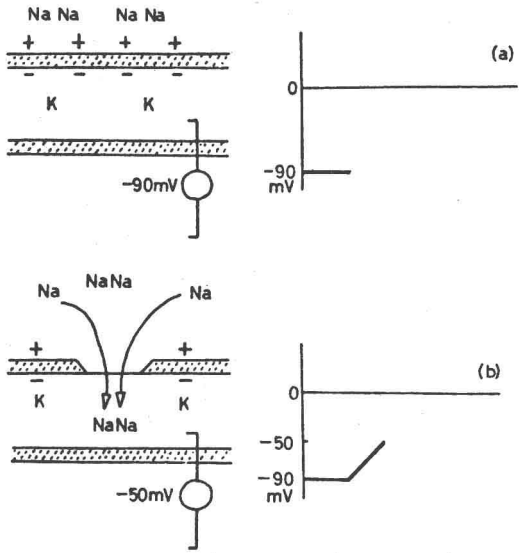

Fig. 3 The sequences of events during depolarization and repolarization.
Fig. 3a. The resting, polarized cell.
Fig. 3b. Acetylcholine causes a local reduction in the membrane potential. Sodium ions begin to flow into the cell.

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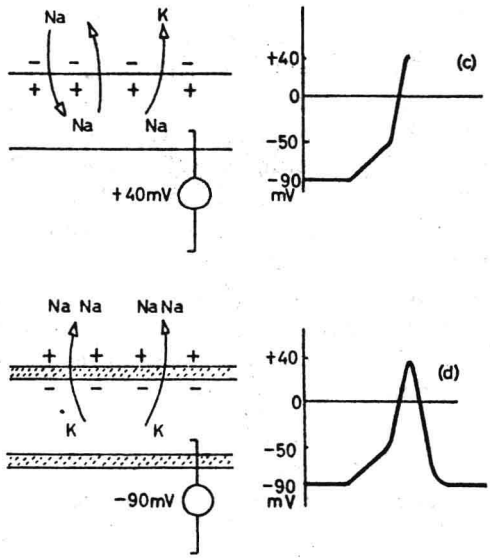

Fig. 3. The sequences of events during depolarization and repolarization.
Fig. 3c. Accumulation of positively charged ions within the cell makes the interior of the fibre locally positive. 'Depolarization'.
Fig. 3d. Sodium ions are expelled from the cell by an active pump mechanism. Repolarization.

In other words, it is polarized. The cellular fluid contains a high concentration of potassium (157 mEq l^{-1}) and a low concentration of sodium (14 mEq l^{-1}). The arrival of acetylcholine induces a reduction in the membrane potential to about —50 mV (Fig. 3b). This increases the permeability of the membrane to the positively charged sodium ions which flow into the cell. The inside of the cell now carries a positive charge (+40 mV). It is depolarized (Fig. 3c). Thus, partial depolarization by acetylcholine at the end plate induces a self-generating wave of depolarization which spreads over the muscle membrane.

During this time the muscle fibre contracts. Before the fibre can respond again, the muscle cells must be repolarized. This occurs when the acetylcholine is broken down by an enzyme cholinesterase which is normally present at the neuromuscular junctiton. The electrolyte constituents of the cell are returned to their resting concentrations by an active pump mechanism (Fig. 3d).

The contractile element of the *muscle fibre* is a combined protein actomycin. The energy for contraction is derived from the breakdown of adenosine triphosphate (ATP). At rest, the actomycin and ATP are firmly bonded together. Depolarization of the muscle membrane is accompanied by a dissolution of this bond, breakdown of ATP, and shortening of the fibre. The muscle fibre is composed of parallel interdigitating filaments. The effect of depolarization is to make these filaments slide together, shortening the overall length of the fibre. While an essential feature of the muscle membrane is its variable permeability to electrolytes, it is for all practical purposes impermeable to the muscle cell enzymes. The appearance of increased amounts of glutamic oxaloacetic transaminase, aldolase, or creatine kinase in the circulating blood, is indicative of primary muscle disease.

DISORDERS OF MOTOR FUNCTION

Disorders of motor function are manifested by three main groups of symptoms:

weakness;
incoordination and involuntary movements;
altered tone.

Weakness

Weakness results from an interruption in the motor pathway at any level, affecting the upper or lower motor neutron, the neuromuscular junction or the muscle.

(1) Upper motor neuron lesions

An upper motor neuron lesion causes weakness of willed movements and it releases irrelevant patterns of muscular tone (spasticity). In health, this hypertonicity is inhibited by the influence of the corticoreticular fibres. The distribution of hypertonic muscles in a 'spastic' limb is not a consequence of haphazard neuronal activity. The extended legs and the flexed adducted arms represent the re-emergence of primitive supporting postures which had been modified by subsequent neuronal maturation.

The weakness is likely to affect movements rather than

individual muscles. Thus, a muscle may be weak or paralysed for the execution of one movement but retain normal power when used in a different movement, e.g. there may be paralysis of voluntary wrist extension, while the same muscles act powerfully as synergists when the patient clenches his fist. Fine, highly differentiated movements, e.g. digital manipulation, are likely to be more severely affected than coarse proximal movements. Muscle tone is increased. The tendon reflexes are brisk and clonus may be present. The plantar reflexes, if involved, are extensor. Muscle wasting occurs only after a delay and is due to disuse. There is no fasciculation.

A destructive or compressive lesion at any level in the brain or spinal cord may be responsible.

Cortical lesions cause contralateral paralysis. Due to the dispersion of the cortical representation a discrete lesion in the motor cortex will give rise to a circumscribed paresis.

Lesions in the *internal capsule* (a common site for cerebral haemorrhage) are likely to cause a complete contralateral hemiparesis as the descending fibres here are grouped closely together.

Brain-stem lesions often affect both corticospinal tracts. If sited above the mid pons they may cause a pseudobulbar palsy. This consists of a spastic weakness of the facial muscles, the muscles of mastication, the tongue and palate. The jaw jerk is increased, the tongue lies stiffly in the floor of the mouth, and there is a spastic dysarthria. Motiveless crying or laughter frequently accompanies a pseudobulbar palsy.

A lesion of the corticospinal tract below the pyramidal decussation gives rise to an ipsilateral spastic paralysis. See Brown-Séquard syndrome (p. 55).

(2) Lower motor neuron lesions

The weakness is likely to involve specific muscles or groups of muscles for all movements. Muscle tone is decreased. The tendon reflexes are depressed or lost early in the disease. The plantar reflexes, if present, are flexor. Muscle wasting occurs early and is prominent. Fasciculation may occur.

Fasciculation is the brief subcutaneous rippling movement seen over the muscle bellies. It is caused by momentary shock-like contractions of the muscle fibres comprising one motor unit.

Disease of the nerve cell body or the proximal part of the axon generates abnormal motor impulses which activate these fibres.

Fibrillation is the spontaneous contraction of single muscle fibres. It is an electromyographic diagnosis and cannot be recognized clinically.

Lower motor neuron signs are likely to be found at the site of any lesion in the brain-stem or spinal cord, combined with upper neuron signs below the level of the lesion. Lesions of the anterior horn cells, nerve roots or peripheral nerves cause isolated lower neuron lesions with a specific distribution. Nerve conduction studies and electromyography may be of assistance in locating the lesion.

Symmetrical, peripheral, lower neuron weakness is described in the section on the neuropathies. Isolated lower neuron weakness is most commonly due to local compression. Neurotropic virus infeetion and exposure to toxic chemicals are less frequently responsible.

(3) Neuromuscular junction

Myasthenia gravis is the only spontaneously occurring example of a disorder at this site. The outstanding feature is a rapid loss of power on sustained or repeated contraction of the muscle, which may be restored to normal by the administration of anti-cholinesterase drugs.

Affected muscles are hypotonic, yet the tendon reflexes are normal or brisk. Wasting is a late feature and fasciculation only occurs as a result of over-treatment.

(4) Muscle

Primary muscle disease often causes symmetrical weakness, affecting the proximal muscles more than the distal ones. Wasting occurs, but is usually less prominent than the weakness, and tendon reflexes are diminished only in proportion to the wasting. Fasciculation does not occur except in thyrotoxic myopathy (p. 201). Electromyography is usually diagnostic. Serum muscle enzymes are raised if the process is active. In the group of inherited muscle disorders (the muscular dystrophies) a family history may be obtained.

Co-ordinated movement

The performance of an unfamiliar manual task requires conscious effort. The initial attempts are slow, stiff and imprecise and fatigue rapidly. With practice these defects are reversed and the action is absorbed into the individual's programme of motor skills. Such a learning process probably involves physical changes in the neurons concerned so that the flow of impulses along a specific pathway is facilitated. Repetition enhances the facilitation.

Fully co-ordinated movements depend on:

1. The cerebral cortex for the organization and analysis of willed movements.
2. The basal ganglia for the semi-voluntary and postural patterned movements.
3. The motor system for the execution of movement.
4. The sensory system to provide constant inflow of information regarding posture, orientation and position of the limbs.
5. The cerebellum to co-ordinate the sensory input. It is then matched against the cortical pattern of required activity and the appropriate modifications are made through the extrapyramidal system (Fig. 2).

(1) Cerebral cortex

The conscious ordering of movement is sometimes called praxis. It depends on the individual's ability to retain the pattern and sequence of a required movement and to translate instructions or intentions into effect. The seat of this cortical organization is in the dominant parietal lobe, from where there are subcortical connections with the motor cortex. The control of cortically organized movements crosses to the non-dominant hemisphere via the corpus callosum.

Disorders of consciously organized patterns of movement are termed apraxias. There is no paralysis, sensory disturbance or ataxia, and the patient fully understands the nature of his task. It is as though his 'memory' for organizing the correct sequence of movements were lost. Movements are performed awkwardly and the patient is often unable to gesture or use simple mechanical instruments such as scissors. Apraxia is closely related to some forms of motor dysphasia.

(2) Basal ganglia

The basal ganglia are nuclear masses buried deeply in the white matter of the cerebral cortex. They form the anterior and lateral relations of the thalamus from which they are separated by the internal capsule (Figs 4, 4a, 4b). Afferent fibres to the basal ganglia come from the motor and supplementary motor areas of the cortex; from the parietal lobe; from the auditory and visual cortex; from the cerebellum and so indirectly from the vestibular

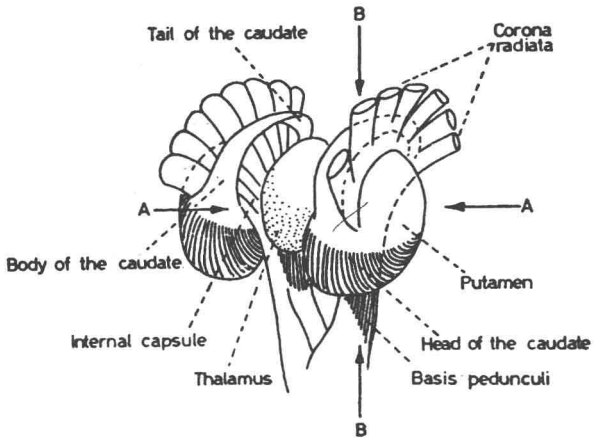

Fig. 4. The basal ganglia and adjacent structures seen obliquely from the front.

system, the postural sensing mechanism of the limbs and the neck muscles.

Both voluntary stimuli and postural reflexes modify the activity of the basal ganglia. Some influences facilitate and some are inhibitory. Facilitatory influences and the outflow from the basal ganglia are mediated by acetylcholine. The substantia nigra has an inhibitory effect which is mediated by dopamine. The major outflow from the basal ganglia is from the globus pallidus to the ventro-lateral nucleus of the thalamus and thence to the reticular formation. At every level there are feedback pathways to those above so that the output is constantly modified.

There are contributions to the pyramidal tract, but the main extrapyramidal outflow is by the reticulospinal and rubrospinal

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