Neurological Emergencies in Medical Practice:

A Handbook for the Non-Specialist

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NEUROLOGICAL EMERGENCIES IN MEDICAL PRACTICE: A Handbook for the Non-specialist

The purpose of this book is to provide a concise, symptom-oriented text on the diagnosis and management of neurological emergencies for the front-line medical practitioner. This might be a family doctor or a hospital doctor working in an accident and emergency department, or whoever first treats the patient. Written by neurologists and neurosurgeons from Britain and the USA who have daily first-hand experience of the subjects they discuss, the aim of the book is to be essentially practical and it should be of immediate value to a wide range of clinicians.

Edited by David Bowsher, Reader in the Department of Neurological Science at the University of Liverpool.

Preface

Neurology has a reputation for being 'difficult' among the majority of doctors who do not practise it in one of its medical or surgical forms. Whether this is because the time devoted to it in the medical curriculum is inadequate, or because (God forbid!) its practitioners behave like High Priests exercising a mystery beyond the ken of lesser mortals must be left to others to decide. Naturally, the editor and authors of this book incline to the former explanation

In addition to a perhaps inadequate early training, neurological/neurosurgical cases are relatively rare among the mass of pathology encountered by the average community or hospital doctor in his or her daily experience. For example, many general practitioners recount that they have *never* seen a case of 'thalamic syndrome'; yet this author currently has over 40 attending his clinic — which probably means that it is commoner than motor neurone disease (amyotrophic lateral sclerosis).

This small book, which will hopefully fit into the glovebox of a car if not into the pocket, has been conceived in order to demystify and explain those many and various neurological emergencies across which any doctor may come any day. What we have set out to do is, first, to describe how to recognise/diagnose the various conditions, and secondly how to deal with them.

So this is essentially a practical manual. If you think any part of it could be improved, please write and tell us how!

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How to Recognise and Examine a Neurological Emergency

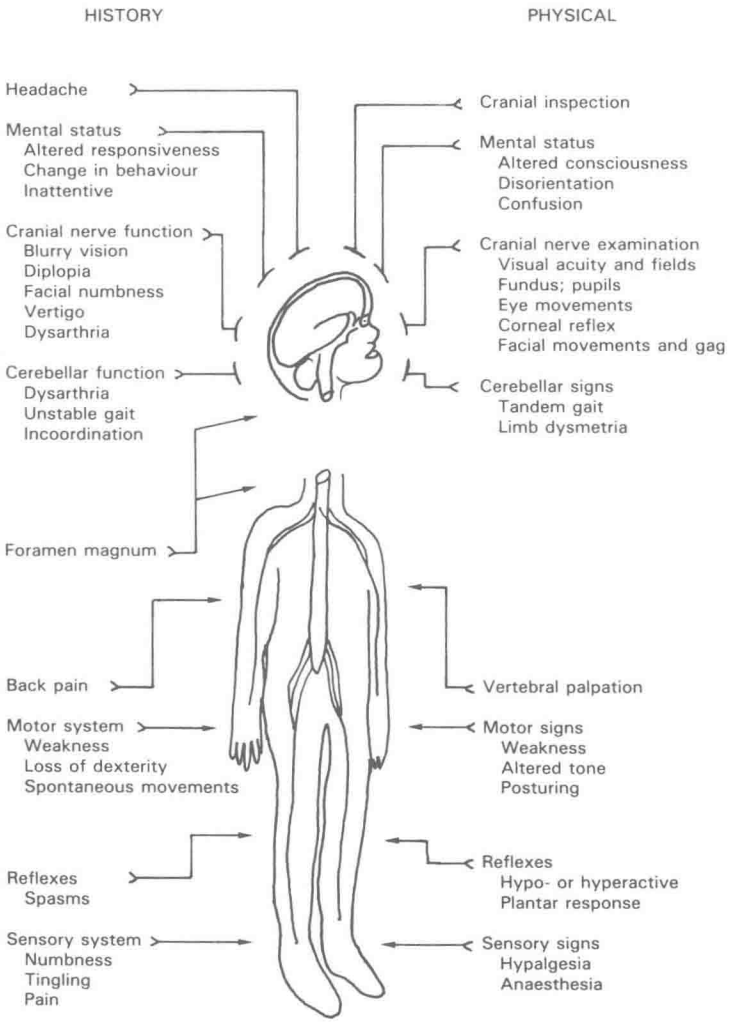
John B. Selhorst

INTRODUCTION

The relevance of a careful neurological assessment is evident in population-based statistics. The prevalence rate for diseases affecting the nervous system in the United States is 9.5%, and new cases are encountered at an annual incidence of 2.5% (Kurtzke, 1984). More importantly, nearly 2% of these cases are acutely disabling and potentially very serious illnesses. Thus, the general practitioner should regularly expect to be confronted with acute neurological disorders.

Not infrequently the non-neurologist's recollections of neuroanatomical pathways and eponymic clinical signs are vague. A reliable and orderly clinical method to classify a neurological illness and begin its management is needed. To tackle an evolving neurological emergency, the time-tested methodology to divide and conquer is employed and accomplished in the following steps. First, the presence of a neurological disorder is acknowledged. Secondly, the process is assigned to either the central or peripheral nervous system or both and localised within the anatomical subdivisions of these systems. Thirdly, the pathology is characterised by the temporal evolution, type and distribution of symptoms and signs. Fourthly, the site and character of the disturbance are confirmed by the neurological examination and appropriately selected diagnostic studies. Finally, the accuracy of this estimation is verified by the response to treatment and course of the illness. The following commentary details how these specific steps are achieved, and an outline of the orderly process is illustrated in the accompanying figure (Figure 1.1).

Figure 1.1: Schematic history and physical for neurologic emergency



THE HISTORY

The clinician's recognition of a neurological disorder begins with the personal introduction. No single moment yields more useful information to the neurological assessment. The chief functions of the nervous system — cognition, locomotion and sensory detection — are immediately evident for inspection. For example, the patient with acute pain or crippled movements is readily apparent. Because the nervous system is the control centre for many complex and highly developed functions, close inspection commonly discloses an abnormality. The mental status examination commences as the physician introduces him- or herself and verifies the identity of the patient. To relax patients and better understand the personal significance of their complaints, it is useful to inquire about their occupation, e.g. roofer, salesman, housewife, secretary. The level of attention, conveyance of affect, choice of language and lucidity of verbal expression are noted and should be directly and further pursued if other than normal. A mental pause to assess the appropriateness of the patient's appearance, behaviour and comfort is rewarding in providing clues to a cerebral malfunction.

After the introduction, the primary complaint is sought. The earnest clinician listens critically to the patient's concerns and choice of words. Further questioning as to just how the symptom complex affects the patient's customary lifestyle and habits often assists in clarifying the presence and magnitude of sensory disturbances, motor deficits or intellectual impairment. Thus, an acute illness striking the nervous system is readily identified.

Next, by employing a series of skilful questions, the clinician establishes which part of the nervous system is disturbed. Attention is especially given to the principal location and distribution of pain as well as the functional complaints. A useful anatomical landmark for the assignment of symptoms is the foramen magnum. This is because the nature of the diseases affecting the nervous system above and below this opening in the skull and the subsequent choice of diagnostic tests to confirm the neurological disorder are so different. The following exposition pertains to symptoms of disorders above the foramen magnum.

Headache results from the stimulation of pain fibres found either in the scalp or contained in the meninges and its blood vessels. Most headaches involving the scalp are due to muscle contraction or migraine-associated changes in the blood vessels. These conditions are emergent in so far as they demand relief. Exceptions among

extracranial headaches are those conditions associated with inflammation or arteritis. Intracranial processes causing acute headache are meningeal infections or disorders resulting in sudden stretching or compression of the meninges such as haemorrhage or hydrocephalus. The headache is commonly aching in quality and either focal or generalised. Pain in the posterior neck is an additional complaint of patients with cerebellar or lower brainstem disease. Intracranial headaches are ordinarily constant or progressive over time, which distinguishes them from the pain-free, episodic intervals that are common with migraine and tension headaches. Vomiting is a less specific sign of intracranial disease but occurs in patients with acute processes involving an increase in pressure or inflammation extending into the ventricular cavity. Presumably, vomiting results from stimulation of the area postrema on the floor of the fourth ventricle. Photophobia is another non-specific symptom that occurs in patients with inflammation of the meninges.

Clearly, neurological symptoms due to intracranial disease are confined to the cerebral hemispheres, brainstem, cerbellum and cranial nerves. A principal interconnecting system is the reticular activating system (RAS) which is compromised in many acute neurological emergencies. The integrity of this system is necessary to activate the cerebral cortex and maintain wakefulness. Altered levels of consciousness occur with a bilateral or diffuse anatomical physiological or metabolic disturbance (Plum and Posner, 1980). The impairment involves either the RAS in the upper pons or midbrain or their projections to the thalamus and the cerebral cortex. Because of the potential gravity for serious neurological disability or death, the mildest form of altered arousal demands attention and necessitates an immediate explanation. Depending on the zone and severity of involvement, disorders of the cerebral cortex affect the quality of mental function and are reflected in altered behaviour, poor concentration, flat affect and poor memory, and in inability to calculate, solve problems, recognise patterns or perform complex motor tasks. Abnormalities in language point to an affliction over the lateral portion of the dominant hemisphere. The general practitioner need not know precisely where or why these symptoms are attributed to the encephalon, but only that they are. When the examiner determines that the patient has an altered level of alertness or disordered thinking, further history from a reliable companion or relative should be sought.

With respect to the motor system, several remarks are appropriate concerning intracranial lesions. Disturbances in the

motor cortex or its pathways descending through the brainstem result in paralysis or various degrees of weakness, slowness and clumsiness of the contralateral lower face and limbs. Acute structural lesions of the basal ganglia are infrequently manifested by the symptoms of a chronic extra-pyramidal disorder. Rather, compression of the contiguous internal capsule results in a hemiplegia or hemiparesis. Toxic reactions, especially to drugs, sometimes present as spontaneous, involuntary movements, e.g. dystonic contractions of muscles, sudden single (myoclonic) jerks and choreiform movements. Acute lesions of the cerebellum are uncommon and are divided into those involving the hemisphere and the midline or vermis. Acute structural lesions of the cerebellar hemispheres result mostly from infarction. Patients experience dysarthria and poor coordination of the ipsilateral limbs. Conversely, acute lesions of the cerebellar vermis, such as occur with hypertensive haemorrhage, produce a sudden loss in control of the paravertebral muscles and vestibular pathways responsible for an unstable posture and gait. These patients present in a wheelchair or on a stretcher because they are able neither to stand nor to walk, and often are unable to sit up without support. To the unsuspecting examiner, there is a surprising absence of weakness, but, if sought, incoordination of the lower limbs is often present. Thus, in every neurological assessment, an important guideline is to determine whether or not the patient can walk, and if not why not.

Sensory symptoms due to intracranial disease are unlikely features of acute neurological disorders. A reduction or alteration in sensation of the limbs suggests involvement of the contralateral brainstem, thalamus, sensory cortex or their intervening connections. Generally, patients complain of tingling or numbness and, infrequently, pain or loss of thermal sensitivity.

Although isolated cranial nerve lesions occur, it is most important to process the localisation to match acute cranial nerve deficits to long tract motor and sensory deficits that appear with lesions within the brainstem. The cranial nerves subserve both the somatic and accessory respiratory muscles and the primary and special sensory functions of the cranium. The somatic muscles are those of the eyes and tongue, and symptoms include ptosis, double vision, dysarthria and dysphagia. The accessory muscles of respiration are involved in jaw, facial, pharyngeal, laryngeal and head movements. Weakness results in ocular exposure, hoarseness or impaired chewing, sucking, swallowing and head-turning.

Numbness of the face above the eyes and tip of the nose

corresponds to the first division of the trigeminal nerve, over the cheeks and upper lip to the second division, and along the mandible and lower lip to the third division. Disorders of the special senses result in loss of vision, hearing, balance, smell or taste. Acute afflictions of independent cranial nerves are uncommon. Loss of vision is the most serious. Usually the patient complains of blurry and dim vision with compromise of the retina or optic nerve. Homonymous impairment or disturbances such as bright flashes or geometric figures in the visual field imply a structural or physiological alteration in the visual pathways behind the optic chiasm and, usually, in the opposite posterior hemisphere. Because auditory pathways are composed nearly equally of crossed and uncrossed fibres, they are rarely clinically affected by focal neurological deficits. Sudden loss of balance due to a vestibular dysfunction is a frightening experience for many patients. The abrupt sensation of spinning or vertigo is regularly accompanied by nausea or vomiting and aggravated by any change in the position of the head. This disabling situation prompts the patient to think that a stroke is occurring. Disorders of smell and taste are not those of acute neurological disease.

Symptoms below the foramen magnum pertain to the spinal cord, peripheral nervous system and muscle. Recognition of impending spinal cord malfunction, which is not uncommon among patients with metastatic disease, is of critical importance. Vertebral pain results from infection or neoplastic invasion that may eventually compress the spinal cord. Symptoms at the thoracic and lumbar level include weakness and sensory loss of the lower limbs and trunk. With partial lesions, paresis occurs in the ipsilateral extremity, and altered sensation involves the opposite limb. Disturbances of the cervical cord result in additional involvement of the upper extremities. Loss of bladder control or acute retention and distension of the bladder may also occur. Complaints pertaining to the inability to evacuate the bowels or to develop an erection are infrequent.

Acute disorders of the peripheral nerves are characterised by distal motor and sensory complaints. There are several explanations for the distal distribution of these symptoms. Over the trunk of a nerve, there is an accumulative effect on altered conduction due to toxic or inflammatory conditions. Thus, the longer the nerve, the greater the disturbance in conduction and sensory perception. In addition, the small muscles of the distal limbs carry out intricate movements that require patterns of precisely timed discharges. Thus, movement of the smaller, distal muscles is more sensitive to the loss of motor neurones or their axons. Early symptoms of motor

dysfunction include stumbling, falling and loss of grip and dexterity. Not infrequently, the paresis encountered in acute neurological emergencies is more profound and results in the inability to walk, stand, or raise the arm. Conversely, the proximal muscles are larger and more actively engaged in tonic contractions that require the expenditure of a large degree of energy to overcome the weight of each limb. Accordingly, diseases of muscle are characterised by proximal weakness. Patients complain about their inability to rise from a chair unassisted, climb stairways, reach overhead shelves or comb their hair. When weakness extends to the paravertebral muscles, patients discover that they are unable to roll over. Sensory disturbances of peripheral nerve disease are also distal. These include numbness and tingling and painful or uncomfortable sensations with stimulation of the affected area. Sharp pain radiating along the course of a peripheral nerve also occurs.

Having assigned the patient's symptoms to areas above or below the foramen magnum and their respective subcompartments, the examiner needs to define the temporal profile of symptoms. This is useful in characterising the underlying pathological process. For example, traumatic and vascular disorders are usually sudden in onset, whereas infections and metabolic and toxic diseases often have a subacute evolution of symptoms. An exact description of where and what the patient was doing at the precise moment of the onset of symptoms is very helpful in defining the abruptness of the neurological event and assisting the patient in recalling any other concurrent symptoms or physiological stresses. Having the patient relate just how the motor, sensory or cognitive malfunction impaired his activity is especially helpful in discerning the magnitude of the symptom.

Additional features in the history assist in estimating the underlying pathology. The occurrence of pain is suggestive of expansion or compression of neural structures or irritation of free nerve endings by inflammation. Episodic symptoms are observed in patients with the neuronal bursts of a seizure or alterations in circulation occurring with vascular disorders. Bilateral and diffuse symptoms are encountered with intoxication and metabolic disturbances of the nervous system.

The history accompanying acute neurological deficits cannot be stressed enough. Frequently, the historical data base constitutes the largest component of an accurate clinical diagnosis. Attention to the foregoing outline should contribute to the clinical method of recognition, localisation and pathological estimation.

THE EXAMINATION

In acute neurological emergencies, the vital signs are of critical importance. A slow pulse rate in a patient with altered consciousness suggests a dangerously high elevation in intracranial pressure. Altered cardiac rhythms are possibly a telling point in a patient with a focal neurological deficit and cardiogenic embolus. Because of loss of autoregulation of cerebral blood flow with structural lesions of the brain, the need to immediately recognise hypotension in patients with neurological disease cannot be over-emphasised. Perfusion of the focally impaired circulation is directly related to the mean arterial pressure. However, the presence of hypertension in acute neurological disorders requires careful judgement before the blood pressure is lowered. Not infrequently, increased blood pressure is the systematic response to the need for a higher perfusion pressure in patients with intracranial hypertension or with a focal loss in cerebral autoregulation. The alternately slow, shallow and rapid, deep excursions of Cheyne-Stokes respirations clue the clinician to physiological alterations affecting the influence of the cerebral hemispheres on respiration. The bradypnoea is sometimes so profound that it is confused with a respiratory arrest. The sudden appearance of other signs of respiratory release, such as yawning, sighing or hiccuping, are also possible early symptoms of bihemispheric dysfunction. In many instances, the potential for aspiration in comatose patients necessitates intubation and mechanical ventilation. This common procedure obscures observation of the neurogenic influences on patterns of respiration. Thus, the laboured hyperventilation and apneustic breathing associated with upper and lower pontine pathology and the ataxic respirations of medullary lesions are rarely encountered. In addition to documenting fever, the determination of body temperature is necessary to recognise patients with hypothermia associated with thiamin deficiency. hypoadrenalism, hypothyroidism, hypoglycaemia or drug intoxication.

The neurological examination is often conducted in the course of the general physical examination. The purpose is to confirm the location of the neurological disorder suspected by the history. Beginning with the top of the head and ending at the foot, the examination is divided into the mental status and cranial nerves, which are structures located strictly above the foramen magnum, and the motor, reflex and sensory subsystems, which indicate dysfunction either above or below the foramen magnum. If the

history regarding motor and sensory function is unremarkable, cerebellar function may be evaluated after the cranial nerves.

In many acute neurological emergencies, the importance of a formal declaration regarding the level of consciousness cannot be overemphasised. Thus, very arbitrary terminology is useful. If other than fully alert, the patient is referred to in the following manner: obtunded, if only a single stimulus arouses the patient to sustained attentiveness; stuporous, if continued stimulation is necessary to maintain the patient's responsiveness to the environment; comatose, if vigorous stimulation fails to elicit a purposeful reaction to the stimulus. Because these terms apply to a fluctuating continuum between wakefulness and coma, the stimulus used and the response elicited should be specifically stated. If consciousness is altered, an extensive examination is limited by the patient's inability to cooperate. Therefore, attention is directed towards determining if the altered level of consciousness is due to impaired activity of the cerebral hemispheres or involves the RAS in the upper brainstem. This achieved by careful observation for signs of brainstem dysfunction reflected in the pupils, eye movements, motor posture and power. These signs also assist in establishing a level of structural or physiological dysfunction in the midbrain, pons or medulla. Consequently, these specific components of the examination are described in detail.

An understanding of pupillary signs is dependent upon a working knowledge of the efferent pathways that influence them. The size of the pupillary aperture is determined by a balance of the parasympathetic and sympathetic nervous system. The parasympathetic influence originates in the Edinger-Westphal nucleus of the rostral midbrain. Pupillomotor fibres pass through the oculomotor nerve, synapse in the ciliary region, and continue as the short ciliary nerves that terminate on the sphincter pupillae muscle. Disruption of these fibres results in mydriasis due to uncontested contraction of the dilator muscle and a fixed reaction to light because of paralysis of the sphincter pupillae. To closely observe the reaction of the iris to light, the 20+ lens of a standard ophthalmoscope is sometimes useful. Impairment of the parasympathetic fibres occurs with compression of the third nerve by the uncus of the temporal lobe, which is forced medially by rapidly evolving hemispheric masses. Often ptosis and adduction paresis from denervation of the levator palpebrae and medial rectus muscle are also present. Bilaterally fixed and dilated pupils are an ominous sign of severe compromise of the rostral midbrain.