DECISION MAKING IN ADULT NEUROLOGY

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Leon A. Weisberg, M.D.

Professor of Neurology
Tulane University

Director of Charity Hospital
Neurology and Neurosurgery Service
New Orleans, Louisiana

Richard L. Strub, M.D.

Professor of Neurology
Louisiana State Medical Center
New Orleans, Louisiana

Carlos A. Garcia, M.D.

Professor of Neurology and Pathology
Louisiana State Medical Center
New Orleans, Louisiana

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PREFACE

Traditionally, the best means of teaching clinical medicine has been via a tutorial system. The system is one by which the experienced clinician imparts the skills and the art of his trade to the young physician by evaluating patients at the bedside and in the clinic. Reading textbooks, journal articles, listening to lectures, and attending conferences may help supplement these practical clinical skills with more didactic material, but rarely does such material give insight into the decision making process used by the seasoned practitioner.

This book is one effort to bridge the gap between didactic learning and practical experience. Decision making trees or algorithms are used to help teach the student to think in the same fashion as the experienced clinician. The basic factor that separates the student's approach to a patient's problem from that of the practicing physician's is the clinician's knowledge and experience with the diseases. The student knows the names and basic pathophysiology of the disease, but is as yet unskilled in arriving at the diagnosis given the patient's symptoms and signs.

Leon A. Weisberg, M.D.
Richard L. Strub, M.D.
Carlos A. Garcia, M.D.

NOTICE

Every effort has been made by the authors, the editors, and the publisher of this book to ensure that dosage recommendations are in agreement with standards officially accepted at the time of publication.

However, it is urged that you check the manufacturer's recommendations for dosage especially if the drug to be administered or prescribed is one recently introduced, in which case dosage schedules are changed from time to time; is one you use only infrequently; or is one you have not used for some time.

The Publisher

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INTRODUCTION

The basic principle of the algorithm is that inherent in the diagnostic process is a certain order, which evolves in a logical, step-by-step analysis. As the history unfolds, the experienced physician asks key questions that lead to possible diagnoses. For example, if a patient faints, your first question is, was it observed? If yes, did the observer notice seizure activity? If so, this leads into a diagnostic path different than if the patient had had a simple syncopal attack. If it was a syncopal attack, did it occur in a typical setting for vasovagal syncope, such as from standing in the hot sun or from having blood drawn? If not, then immediately the physician must take the symptom much more seriously and begin to consider cardiac disease and a host of other factors.

Decision making is not always as orderly as we suggest, but we hope that this book will impose some order on the reader's diagnostic approach. To this end, we have tried to cover most of the common clinical problems encountered in the usual practice of neurology. We have also provided selected references in each section, and these can be used to expand on the information presented here.

Many of the algorithms discuss a single symptom or clinical sign (e.g., headache, nystagmus, and so forth), whereas others concern specific disease entities such as multiple sclerosis. The symptom-oriented chapters readily lend themselves to both a diagnostic and treatment approach; however, the disease-oriented chapters emphasize management and give only general diagnostic points.

The student should begin each chapter by reading the short related introductory paragraph on the text page and then proceed to the relevant point on the decision tree. The capital letters that appear within the tree refer to paragraphs of text on the facing page. These paragraphs contain explanatory information that is too cumbersome to include in the algorithm itself.

This book is an introduction to the clinical evaluation and treatment of the most common neurological conditions, but this material must be supplemented by standard text material, articles, and, of course, clinical experience.

NEUROLOGIC LOCALIZATION

Leon D. Weisberg, M.D.

As the initial step in the neurologic localization of a lesion, determine the relation of the lesion to the foramen magnum (above or below). Another consideration is whether the lesion is contiguous with the foramen magnum at the craniocervical junction. Lesions below the foramen magnum usually cause bilateral motor and sensory symptoms involving the arms, trunk, and legs. Patients with craniocervical lesions may present with a bewildering array of symptoms; upper spinal cord dysfunction and lower cranial nerve symptoms are often accompanied by posterior cervical and suboccipital pain. Such lesions include syringomyelia, foramen magnum tumors, and base of the skull lesions (e.g., Arnold-Chiari malformation). Lesions above the foramen magnum may be divided into those located above (supratentorial) and below (infratentorial) the tentorium.

A. Lesions that are contiguous with the foramen magnum may extend into the posterior fossa (involvement of ninth through twelfth cranial nerves) or the cervical spinal cord (upper motor neuron signs, including spasticity, Babinski sign, clonus in the legs and later in the arms, pain and temperature sensation loss below the level of the lesion, and sometimes weakness and wasting in the arms). Lesions involving the spinal cord frequently cause bowel and bladder involvement.

B. Infratentorial structures include the brain stem, cerebellum, and fourth ventricle. Lesions of these structures may be intra-axial (located within the cerebellum or brain stem) or extra-axial (e.g., cerebellopontine angle). Cerebellar lesions originate in the vermis (midline) or hemispheres (lateral). Vermal lesions cause gait and leg ataxia, whereas hemispheric lesions cause unilateral cerebellar dysfunction with ataxia and dysmetria ipsilateral to the lesion. Brain stem lesions may cause unilateral or bilateral, symmetrical or asymmetrical cranial nerve, corticospinal, and cerebellar disturbances. Cerebellopontine angle lesions cause unilateral auditory, vestibular, or facial nerve dysfunction, since these lesions may expand and compress the brain stem and cerebellum.

C. Supratentorial lesions may be lateralized or midline. Lateralized lesions may be cortical (frontal, temporal, parietal, occipital) or subcortical (internal capsule, thalamus, basal ganglia). Cortical lesions may involve the dominat temporal lobe, causing language disorders, or the parietal lobe, causing spatial or constructional impairment. Occipital lesions cause visual disturbances (homonymous hemianopia). Frontal lesions may cause changes in personality and behavior. Seizures are common with cortical lesions. Neurologic signs depend upon the supratentorial region involved. Involvement of the medial frontal lobe may result in leg paresis; lateral convexity lesions cause paresis of the face and arm with sparing of the leg. Subcortical internal capsular lesions may cause dysarthria but not aphasia; such a lesion may cause motor or sensory disturbance with equal involvement of the face, arm, and leg. Midline supratentorial lesions may be intraventricular and may obstruct the lateral or third ventricle; these lesions cause intracranial hypertension without localizing signs. Extraventricular midline juxtasellar lesions produce endocrine and visual symptoms.

D. Lesions below the foramen magnum may involve the spinal cord (myelopathy) or spinal root (radiculopathy) and cause sensory and motor disturbances. Spinal root lesions are most common in the cervical and lumbar regions. These lesions usually cause neck or back pain. The distribution of motor, reflex, and sensory abnormalities depends upon the specific root involved. If the spinal cord is involved, there is usually sphincter dysfunction with motor and sensory abnormalities below the lesion level (cervical lesions may involve the arms and legs; thoracic lesions involve legs only). Lesions involving the conus medullaris (lowest portion of the spinal cord) cause sphincter dysfunction with sensory disturbances that involve the sacral region (bottom of feet and the rectal or saddle area). Cauda equina lesions involve multiple lumbar and sacral spinal roots and cause weakness and wasting in the legs with a radicular pattern of sensory impairment. If there are motor findings (with fasciculations and distal weakness), consider anterior horn cell disease. If there is sensory and motor involvement with a distal stocking-glove distribution, consider peripheral neuropathy. If weakness is proximal with normal sensation and normal reflexes, consider neuromuscular junction or muscle disease.

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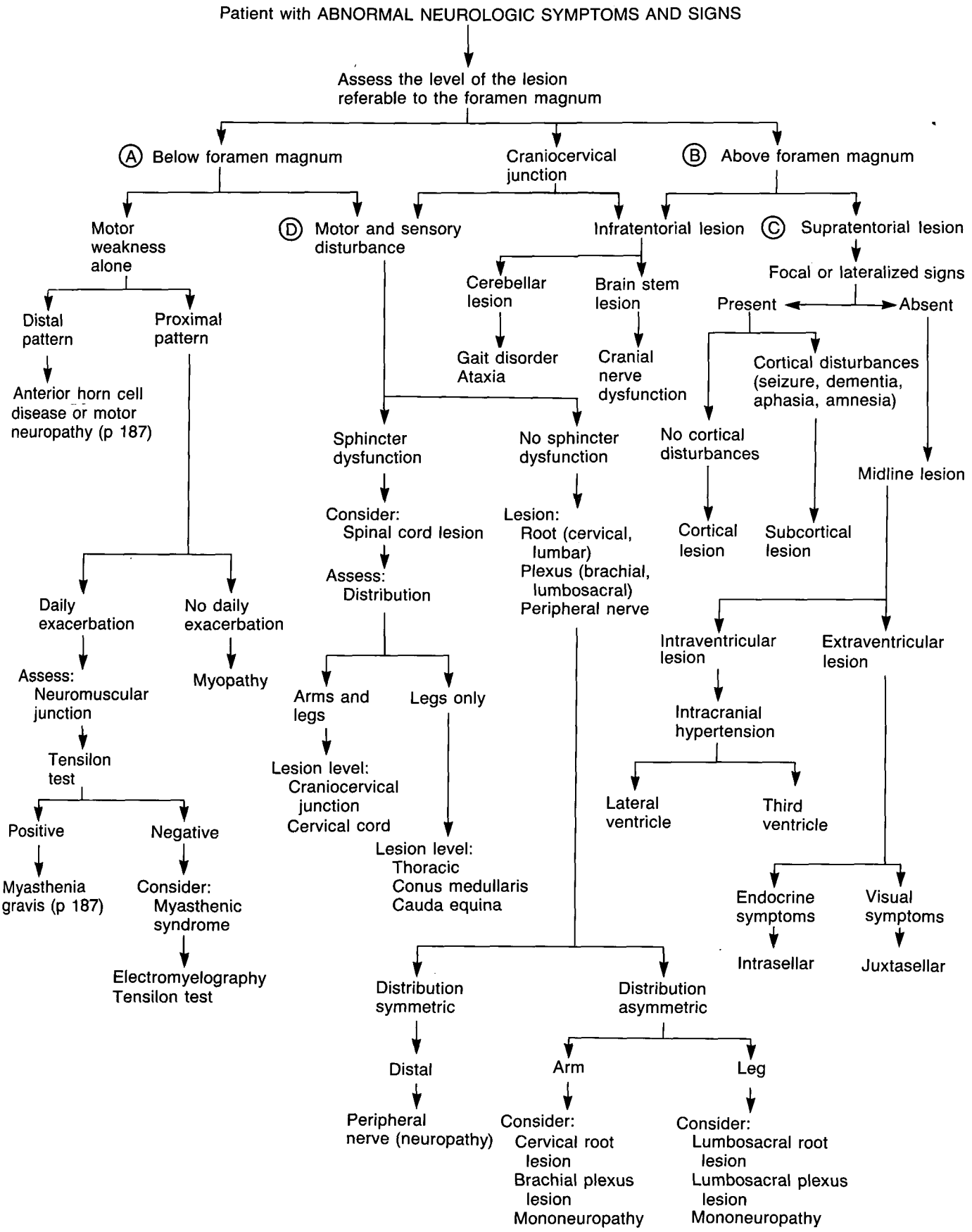
Patient with ABNORMAL NEUROLOGIC SYMPTOMS AND SIGNS
Assess the level of the lesion referable to the foramen magnum
Ⓐ Below foramen magnum
Craniocervical junction
Ⓑ Above foramen magnum
Motor weakness alone
Ⓓ Motor and sensory disturbance
Infratentorial lesion
Ⓒ Supratentorial lesion
Distal pattern
Proximal pattern
Cerebellar lesion
Brain stem lesion
Focal or lateralized signs
Present
Absent
Anterior horn cell disease or motor neuropathy (p 187)
Gait disorder Ataxia
Cranial nerve dysfunction
Cortical disturbances (seizure, dementia, aphasia, amnesia)
Sphincter dysfunction
No sphincter dysfunction
No cortical disturbances
Midline lesion
Consider: Spinal cord lesion
Lesion: Root (cervical, lumbar) Plexus (brachial, lumbosacral) Peripheral nerve
Cortical lesion
Subcortical lesion
Daily exacerbation
No daily exacerbation
Assess: Distribution
Intraventricular lesion
Extraventricular lesion
Assess: Neuromuscular junction
Myopathy
Arms and legs
Legs only
Intracranial hypertension
Tensilon test
Lesion level: Craniocervical junction Cervical cord
Lateral ventricle
Third ventricle
Positive
Negative
Lesion level: Thoracic Conus medullaris Cauda equina
Endocrine symptoms
Visual symptoms
Myasthenia gravis (p 187)
Consider: Myasthenic syndrome
Intrasellar
Juxtasellar
Electromyelography Tensilon test
Distribution symmetric
Distribution asymmetric
Distal
Arm
Leg
Peripheral nerve (neuropathy)
Consider: Cervical root lesion Brachial plexus lesion Mononeuropathy
Consider: Lumbosacral root lesion Lumbosacral plexus lesion Mononeuropathy

ONSET OF NEUROLOGIC DISORDERS

Leon D. Weisberg, M.D.

If the onset is sudden, consider electrical (seizures with paroxysmal neural discharge, demyelinating disorders with conduction block, migraine with spreading cortical depression) or vascular conditions (ischemia, hemorrhage). If the onset is progressive, consider atrophic (degenerative) disorders or a mass lesion (intracranial, intraspinal). Mass lesions usually cause symptoms by direct neural compression or local tissue infiltration, and rarely by vascular mechanisms (peritumoral hemorrhage, ischemia due to blood vessel compression by tumor).

A. The presence of "warning" symptoms is important. In cases of syncope these may be nonfocal (dizziness, weakness, visual blurring); focal symptoms (motor, sensory, visual) occur in migraine, transient ischemic attacks (TIAs), and partial seizures. In TIAs, which may herald the onset of a fixed stroke syndrome, focal symptoms are maximal at the onset with no spread or progression; complete resolution usually occurs within 30 minutes, but symptoms may persist for 24 hours. In partial (focal) seizures, there may be "Jacksonian" spread, an orderly progressive spread of motor or sensory abnormality over the limbs, trunk, or face within a several-minute interval as excitatory electrical activity rapidly spreads. There may also be "Todd's postictal paralysis," which may persist for 24 hours. In migraine, neurologic disturbances may evolve during a 10 to 30 minute interval. The characteristic visual disturbances spread from the periphery to the center of visual fixation; visual symptoms are followed by a contralateral throbbing headache. The evolution of symptoms is slower in migraine (owing to spreading cortical depressive activity) than in seizures, in which symptoms evolve more rapidly owing to cortical excitatory activity.

B. If fixed focal signs develop suddenly, consider vascular or demyelinating disease. In patients with suspected vascular disease, a focal deficit without a warning TIA or subsequent deterioration suggests intracerebral hemorrhage. In cerebral infarction, warning TIAs commonly occur; deterioration may occur after the onset of a fixed deficit (progressing stroke); this is more common in infarction than in hemorrhage. The onset of a neurologic deficit may be sudden in multiple sclerosis (optic neuritis, transverse myelitis), a demyelinating disorder, frequently accompanied by symptoms caused by conduction (electrical) block. Rapid stabilization is frequently followed by spontaneous gradual improvement. Less commonly in multiple sclerosis the onset is slow, and the course is progressive deterioration; this simulates the presentation in patients with mass lesions. Although demyelinating disorders usually occur in young patients, cerebrovascular disease occurs in all age groups. Diagnostic studies may be required to differentiate demyelinating from vascular disease (CT or MRI scan, CSF examination, angiography).

C. Paroxysmal disorders in which symptoms resolve rapidly and the patient returns to baseline condition (e.g., TIA, seizure, migraine, narcolepsy, vertigo, drop attacks, syncope) present diagnostic difficulties. Determine whether consciousness is impaired. In narcolepsy recurrent and excessive brief sleep episodes occur; differentiation from seizures is established by electroencephalography and sleep monitoring. In patients who suddenly lose consciousness without abnormal motor activity, consider Stokes-Adams attacks or hypotension; perform cardiac monitoring. In patients with episodic confusion, consider vascular disorders or hypoglycemia (especially in diabetic patients). Certain disorders impair equilibrium. In drop attacks, the patient suddenly falls to the ground; these attacks may occur in children with seizures; the electroencephalogram is abnormal. In older patients who develop drop attacks without loss of consciousness, consider vertebrobasilar TIA or electrolyte disturbances (potassium in periodic paralysis). In vertigo due to vestibular dysfunction, there are sudden attacks of "spinning" and dysequilibrium; the absence of other brain stem or cerebellar symptoms differentiates vestibular dysfunction from vertebrobasilar TIA.

D. In patients with neurologic symptoms of insidious onset, examination findings are most important. If the examination is negative and symptoms have been present for a prolonged time (several years), consider a functional psychiatric disorder. In patients who have abnormal neurologic findings, consider an atrophic-degenerative disorder or mass lesion. With degenerative disorders, the course of subsequent deterioration is slowly progressive. If there is a mass lesion, the course may be characterized by more rapid progression; however, some patients with low grade gliomas or meningiomas may not deteriorate rapidly. In some patients with these types of tumors, seizures may be the only clinical manifestation for many years, and only later does other neurologic deterioration occur.

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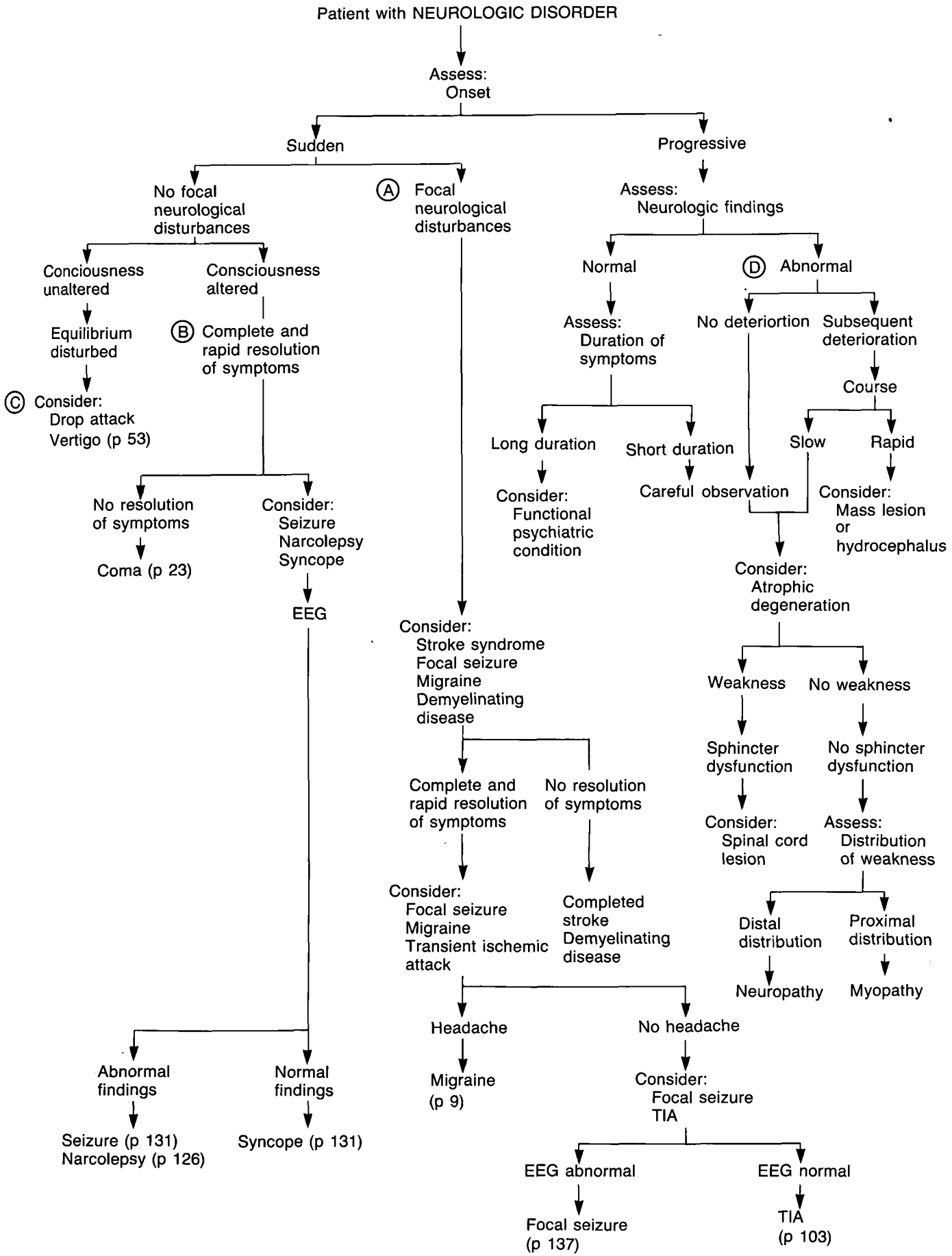

Patient with NEUROLOGIC DISORDER
Assess:
Onset
Sudden
Progressive
No focal neurological disturbances
Ⓐ Focal neurological disturbances
Assess:
Neurologic findings
Conciousness unaltered
Consciousness altered
Normal
Ⓓ Abnormal
Equilibrium disturbed
Ⓑ Complete and rapid resolution of symptoms
Assess:
Duration of symptoms
No deteriortion
Subsequent deterioration
Ⓒ Consider:
Drop attack
Vertigo (p 53)
Course
Long duration
Short duration
Slow
Rapid
No resolution of symptoms
Consider:
Seizure
Narcolepsy
Syncope
Consider:
Functional psychiatric condition
Careful observation
Consider:
Mass lesion or hydrocephalus
Coma (p 23)
EEG
Consider:
Atrophic degeneration
Consider:
Stroke syndrome
Focal seizure
Migraine
Demyelinating disease
Weakness
No weakness
Complete and rapid resolution of symptoms
No resolution of symptoms
Sphincter dysfunction
No sphincter dysfunction
Consider:
Spinal cord lesion
Assess:
Distribution of weakness
Consider:
Focal seizure
Migraine
Transient ischemic attack
Completed stroke
Demyelinating disease
Distal distribution
Proximal distribution
Neuropathy
Myopathy
Abnormal findings
Normal findings
Headache
No headache
Migraine (p 9)
Consider:
Focal seizure
TIA
Seizure (p 131)
Narcolepsy (p 126)
Syncope (p 131)
EEG abnormal
EEG normal
Focal seizure (p 137)
TIA (p 103)

COURSE OF NEUROLOGIC ILLNESS

Leon A. Weisberg, M.D.

Rapid stabilization and spontaneous improvement after the onset of fixed focal deficit are consistent with cerebrovascular disorders (infarction, hemorrhage). After the initial deficit develops in stroke, progression may occur for 24 hours in carotid lesions and for 72 hours in vertebrobasilar territory lesions. Clinical improvement after the stroke is most rapid the first 6 months but may continue for several years.

A. Define stroke by the temporal profile: (1) TIA: episodes last 10 to 30 minutes but may last longer and all focal signs resolve within 24 hours. (2) Reversible ischemic neurologic deficit (RIND): the focal deficit persists longer than 24 hours but resolves completely within several weeks. (3) Minor stroke: 80% functional neurologic recovery occurs within 3 weeks. (4) Major stroke: less complete and slower recovery than in a minor stroke. Patients with spontaneous or hypertensive intracerebral hematomas do not usually develop recurrent hemorrhage; intracranial hemorrhage caused by aneurysms or angiomas may recur or clinical condition may deteriorate after the initial bleeding episode because of ischemia or hydrocephalus.

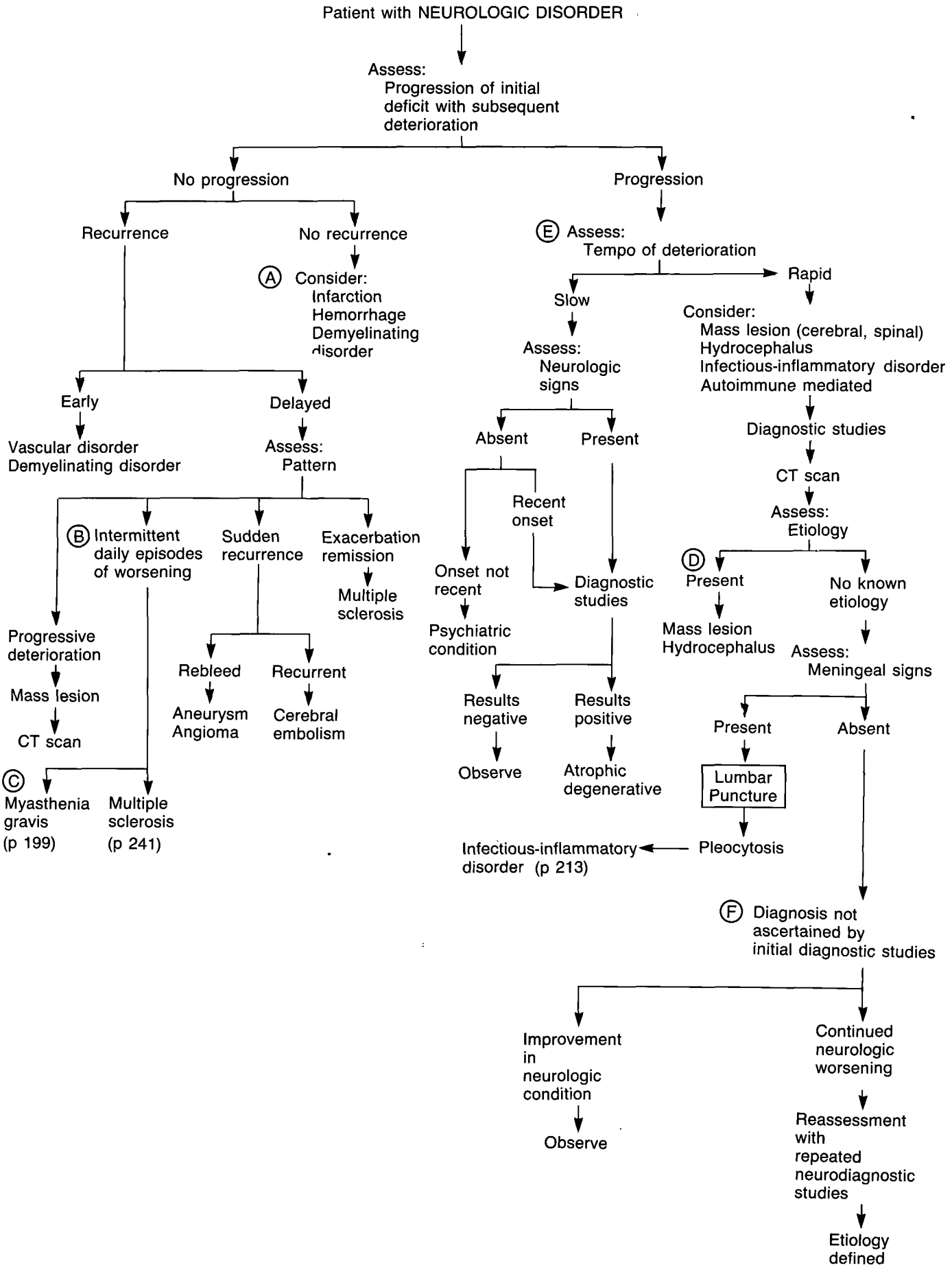
B. The sudden onset of neurologic signs with subsequent improvement is most consistent with cerebrovascular or demyelinating disease. A "triphasic" course characterized by a sudden onset, followed by stabilization and improvement and later by progressive deterioration, is seen with metastases as well as in glioblastoma multiforme and brain abscess. In patients with metastases, hematogenous dissemination of a tumor embolus initially causes infarction; this results in sudden onset and rapid stabilization. Later, as tumor cells multiply, enlargement of the tumor mass and surrounding edema result in neurologic deterioration.

C. Exacerbation of the deficit after the initial onset may occur in several patterns. In myasthenia gravis, periods of neurologic worsening are observed throughout the day; however, between these periods the patient may appear neurologically normal. In patients with a deficit caused by multiple sclerosis (MS) there may be episodes of neurologic worsening lasting days to weeks. During these episodes the patient may develop an exacerbation of preexisting deficit or new patterns of neurologic deficit. Exacerbations may be interspersed between periods of clinical remission. Some patients with MS report worsening of neurologic impairment occurring toward the end of each day, described as "fatigue" or "having no energy." These symptoms may be caused by physiologic conduction block. The history of fluctuation in MS may be similar to that reported by patients with myasthenia gravis.

D. If there is waxing and waning of diffuse neurologic function (for example, confusion alternating with normal consciousness) throughout the day, consider subdural hematoma or another mass lesion, especially if focal signs develop. The development of confusional episodes at night is seen in Alzheimer's dementia (sundown effect). If there is change of focal deficit (for example, a shift from left to right hemiparesis), consider bilateral subdural hematoma, basilar artery occlusion, and recurrent cerebral embolism.

E. Patients may show progressive worsening after initial recognition of a neurologic deficit. If progression is slow and examination shows positive neurologic signs, consider an atrophic-degenerative disorder. If more rapid deterioration occurs, consider a mass lesion. Some patients with mass lesions (gliomas, arteriovenous malformations, meningiomas) initially present with seizures without a neurologic deficit. As the lesion enlarges, a focal deficit may develop. In patients with headache or other neurologic symptoms of longer than one year in duration and no abnormal neurologic signs, neurodiagnostic studies are unlikely to show an underlying structural lesion, even if the patient reports that these symptoms are becoming more severe.

F. A monophasic course with spontaneous improvement suggests a vascular or demyelinating disorder. If the course is biphasic (initial illness with full recovery followed by second phase of neurologic deterioration), consider parainfectious conditions (possibly autoimmune mediated), such as those affecting the peripheral (polyneuropathy of the Guillain-Barré type) or central (demyelination, as in multiple sclerosis or encephalomyelitis) nervous system. These may follow a viral illness or vaccination and appear to result from an inflammatory response of the cell mediated type against peripheral or central myelin. If there is neurological improvement, observe the patient. If there is continued neurologic worsening then repeated neurodiagnostic studies are necessary.


Patient with NEUROLOGIC DISORDER
Assess:
Progression of initial deficit with subsequent deterioration
No progression
Progression
Recurrence
No recurrence
A Consider:
Infarction
Hemorrhage
Demyelinating disorder
Early
Delayed
Vascular disorder
Demyelinating disorder
Assess:
Pattern
B Intermittent daily episodes of worsening
Sudden recurrence
Exacerbation remission
Multiple sclerosis
Progressive deterioration
Mass lesion
CT scan
Rebleed
Recurrent
Aneurysm
Angioma
Cerebral embolism
C
Myasthenia gravis
(p 199)
Multiple sclerosis
(p 241)
E Assess:
Tempo of deterioration
Slow
Rapid
Assess:
Neurologic signs
Consider:
Mass lesion (cerebral, spinal)
Hydrocephalus
Infectious-inflammatory disorder
Autoimmune mediated
Absent
Present
Diagnostic studies
CT scan
Recent onset
Assess:
Etiology
D
Present
No known etiology
Onset not recent
Diagnostic studies
Mass lesion
Hydrocephalus
Psychiatric condition
Assess:
Meningeal signs
Results negative
Results positive
Present
Absent
Observe
Atrophic degenerative
Lumbar Puncture
Pleocytosis
Infectious-inflammatory disorder (p 213)
F Diagnosis not ascertained by initial diagnostic studies
Improvement in neurologic condition
Observe
Continued neurologic worsening
Reassessment with repeated neurodiagnostic studies
Etiology defined

INITIAL HEADACHE

Richard L. Strub, M.D.

Headache is the most common medical symptom. Although usually benign, at times it can represent the first symptom of a serious neurologic disorder. The physician must distinguish the serious from the benign and, having done so, must define the specific type of headache and its etiology.

The history is the most important part of the evaluation of the patient with headache; care must be taken to elucidate not only the character of the pain but also the course of the headache. In addition to the profile of the presenting headache, it is important to distinguish the chronic recurrent headache, which is rarely life threatening, from the first time headache that may reflect serious disease.

A. The patient with a first time headache and positive neurologic findings very likely has intracranial disease, possibly a hemorrhage, meningitis, or acute hydrocephalus. A full neurologic evaluation is indicated.

B. The patient with a sudden headache and completely normal findings on evaluation must be watched carefully. Most neurologists would obtain a computed tomographic (CT) brain scan, and many would perform a spinal puncture. If no clear benign etiology is present, it is prudent to rule out serious disease such as subarachnoid hemorrhage.

C. Many medical illnesses are accompanied by headache, e.g., hypertension, infection, pulmonary failure. Also, a side effect of some medications is headache. Physical strain and psychologic stress can also cause sudden headache.

D. Every headache patient has a first tension, migraine, or cluster headache. Often it is prudent to evaluate the first vascular headache extensively, particularly in those with neurologic symptoms at the onset. In young females, systemic lupus erythematosus is often heralded by a migraine type of headache. Vascular malformations may also present initially with unilateral headache.

E. The temporomandibular joint syndrome is one cause of headache, usually in the temporal region. It is frequently seen after recent dental work or in patients who grit their teeth at night. The joint itself is usually tender to palpation.

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