DISORDERS OF THE NERVOUS SYSTEM

A Primer

ALEXANDER G. REEVES

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Disorders of the Nervous System

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To
Wilbur D. Hagamen,
Teacher Supreme,
model for all who
would teach neural sciences

Preface

THIS BOOK is not comprehensive in scope. It is meant to be a preliminary clinical neurology text for medical students. It is the compilation of eight years' experience teaching introductory neurology to second-year medical students at Dartmouth. General references for further reading are given at the end of each chapter and it is assumed that, as at Dartmouth, faculty will supplement the students' reading with current journal and textual materials when appropriate. A basic course in neural sciences has been a prerequisite and neuropathology and physical diagnosis are taught in parallel. A clerkship on the neurology wards has traditionally followed.

There are two basic divisions in the text. The first six chapters constitute an introduction to neurologic diagnostic concepts. An attempt is made to introduce a basis for the neurologic evaluation and, to a lesser degree, the methods. These can be taught in parallel or subsequently in physical diagnosis courses. The remaining fourteen chapters deal with selected aspects of neurologic symptoms and disease. Well-defined principles are introduced, but there are frequent speculative constructs in neurologic pathophysiology that, it is hoped, will stimulate discussion and further reading.

We are indebted to many people who have assisted in the production of this book: to our neurology residents and colleagues and many generations of medical students who have given incalculable and critical input during evolution of the text and neurology teaching at Dartmouth; to Joan Clifford and Judy Murphy for their secretarial and copyreading expertise; to the Sandoz Foundation and Dr. Craig Burrell for their timely support and advice during the preparation of the manuscript; to Drs. Louis Caplan and Laurence Levitt for their critical and constructive review of the manuscript; and, finally, to Fred Rogers and Edward Quigley and the staff at Year Book for their editorial assistance and patient encouragement.

ALEXANDER G. REEVES

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Part I

Evaluation of the Nervous System

1

General Physical Examination

IN THIS CHAPTER we consider some aspects of the general physical examination that are especially pertinent to neurologic evaluation. In later chapters we discuss other aspects of the neurologic examination and the involvement of specific disease processes and systems.

Vital Signs

Of particular importance is evaluation of the *respiratory rate and pattern* in patients with depressed consciousness. This topic will be elaborated on in the section on the evaluation of coma in chapter 19. Segmental involvement of the brain stem from the diencephalon through the medulla may be associated with characteristic and therefore localizing patterns of respiration at each level of involvement as long as there is *bilateral* dysfunction. Unilateral dysfunction is usually not reflected by respiratory abnormalities. The abnormal patterns of breathing most likely represent loss of higher control of the primary medullary respiratory center. Suppression of medullary function by metabolic or direct mechanical involvement results in apnea; an apneic patient will be asphyxiated if respiratory support is not supplied.

Figure 1-1 is a schematic of respiratory patterns elicited by bilateral lesions at various levels in the brain stem.

Though relatively nonspecific and of little value in localizing, *blood pressure* is frequently elevated considerably above premorbid levels when there is increased intracranial pressure and then drops when intracranial pressure is lowered. Blood pressure drops to very low levels following loss of medullary function; however, for therapeutic purposes severely depressed blood pressure should be assumed secondary to blood loss until proved otherwise. For a person with peripheral nervous system dysfunction (called *peripheral neuropathy*), which may be the result of many factors (i.e., diabetes, alcoholism, or malnutrition), symptomatic hypotension may occur on assuming the erect position (orthostatic hypotension). This can be related to involvement of the peripheral autonomic nervous system with loss of peripheral vasomotor tone. Isolated central or peripheral autonomic failure secondary to drug ingestion and metabolic or degenerative disease processes is also a frequent cause of orthostatic hypotension and other autonomic dysfunction. Orthostatic hypotension can be elicited most easily by having the patient exercise prior to testing; this causes a reactive peripheral vasodilatation that slowly returns to normal when the person is at rest.

The *pulse rate* may be slowed (bradycardia) or hastened (tachycardia) with increased intracranial pressure, and therefore any change must be suspect in a person with central nervous system involvement. Arrhythmias, particularly sinus arrhythmias, and nonspecific ST-T wave changes are frequently seen on electrocardiograms of per-

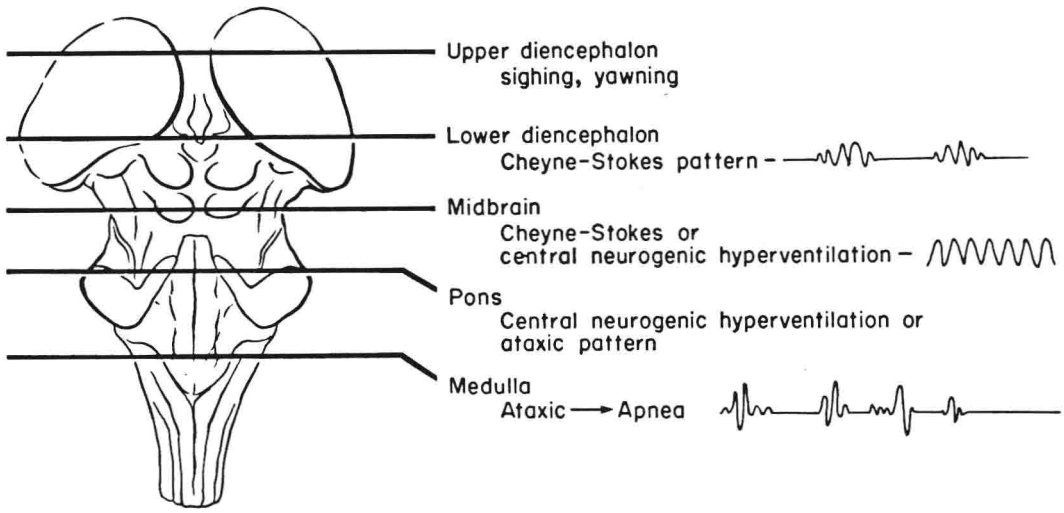

Fig 1–1. — Patterns of breathing associated with various levels of bilateral brain stem involvement (see also chap. 19).

sons who have had either hemorrhagic or ischemic strokes. If auricular fibrillation is present, systemic embolization of thrombus formed in the nonpulsatile left atrium should be considered the likely cause of a stroke.

Head

Changes in the *shape* and *size* of the cranium frequently reflect changes in the intracranial contents.

In children younger than the age of closure of skull sutures, increased intracranial pressure is reflected in widened suture lines that are frequently palpable and quite visible in radiographs. If the pressure is prolonged, a mottled decalcification or beaten silver appearance of the skull and demineralization of the dorsum sellae may appear on x-rays. Bulging of the anterior fontanelle in the erect or seated infant is a reliable sign of increased intracranial pressure or contents. Progressively enlarging ventricles, hydrocephalus, causes enlargement of the skull, which can be observed easily. Early diagnosis of hydrocephalus is possible by measuring the cranial circumference on standard well-baby checkups and comparing it with standard charts. Subdural fluid effusions, usually associated with meningitis and subdural or epidural hematomas, also cause excessive skull enlargement in infants and toddlers who have nonfused suture lines. Premature closure of sutures causes characteristic distortions that should be recognized early because associated restriction of brain expansion may result in neuronal damage and mental retardation (Fig 1–2).

In adults, the shape and size of the cranium are less often revealing. The presence of asymmetric bony prominences contralateral to sensorimotor deficits or in a person with focal seizures suggests an underlying meningioma, a benign tumor that occasionally causes secondary osteoblastic activity in proximate parts of the skull.

In both adults and children who have a history of head trauma or in persons who are stuporous or comatose for unknown reason, the skull should be gingerly palpated for soft-tissue swelling, which suggests head trauma and possible underlying fracture. Ecchymoses around the eyes or over the mastoid region suggest recent head trauma. Natural lymphatic drainage of blood breakdown products in the scalp collects in these areas.

When the head is gingerly rotated from side to side, the brain, which is essentially floating in the subarachnoid cerebrospinal fluid and is tethered to the dura and contig-

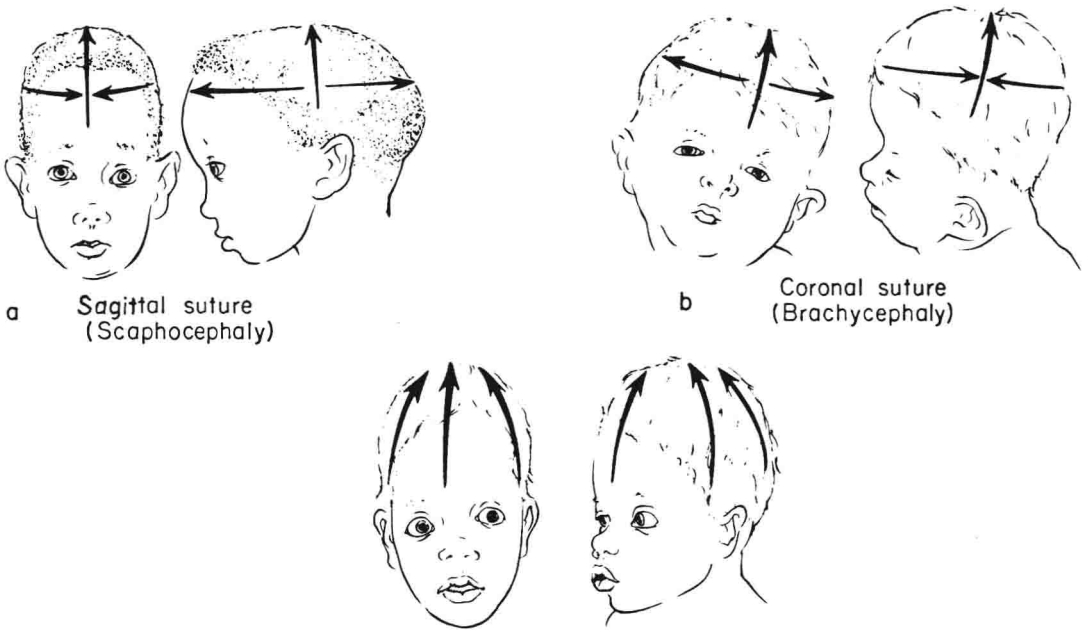

Fig 1–2. – Head shapes associated with premature closure of sutures. **a**, sagittal; **b**, coronal; and **c**, both.

uous skull by cranial nerves, blood vessels, and arachnoid membranes, is relatively resistant to the movement. The movement stretches the cranial nerves, the larger blood vessels, and the dura. Under normal circumstances, no significant discomfort is caused by this stress. However, when the surface blood vessels or dura, which are sensitive to pain, are already swollen from a mass lesion or from inflammatory edema as with migraine or arteritis, pain may be increased or experienced for the first time on shearing. The patient frequently can point to the area of involvement.

Auscultation of the cranium (with the bell of the stethoscope) over the mastoid region, temporal region, forehead, closed eyes, and in bald individuals more extensively is carried out in attempt to identify a vascular bruit arising from an arteriovenous malformation over the brain surface. The bruit is caused by the increased flow in the arteriovenous short-circuit that makes up the malformation.

Physicians do not routinely listen to patients' heads. Indeed, if all the examinations that we have described and will describe were carried out on all patients, it would be difficult to see more than several patients a day. An efficient diagnostic approach demands careful evaluation and utilization of historical data in a problem-oriented fashion. This demands that the physician focus on those parts of the physical examination that are pertinent to the specific problem or problems elicited by history or by basic screening.

Whose head should be auscultated? The person with a history suggesting an arteriovenous malformation is one candidate. Episodic headaches always restricted to the same side of the head are usually migraine; however, most migraine sufferers claim occasional contralateral headaches. Arteriovenous malformation headaches are almost always limited to the side of the head with the abnormality, and for this reason physicians should listen to the heads of patients

who have headaches that always occur on the same side. Most of these patients have migraine, but an occasional one has a malformation. Focal or generalized seizures in a person with unilateral headache suggest brain tumor first but also arteriovenous malformation; this demands auscultation. On occasion the person with arteriovenous malformation hears the bruit, particularly at night when distractions are at a minimum.

The small child or infant with a congenital arteriovenous malformation in the area of the internal cerebral veins and the vein of Galen has a bruit that is audible over the whole head. He may have congestive heart failure from the high flow demands of the shunt, and also an enlarging head, the result of a communicating hydrocephalus. This is caused by the high pressure in the sagittal sinus and therefore increased resistance to absorption of cerebrospinal fluid through the arachnoid granulations. Compression of the aqueduct of Sylvius by the vein of Galen enlargement may also be the cause of the hydrocephalus.

The infant with meningitis and diffuse cerebral vasodilatation and the infant or child with severe anemia may have diffuse cranial bruits caused by high flow through the cerebral or diploic vasculature. These bruits are usually inaudible in the older child or adult whose skull is thicker and thus dampens the sound.

Eyes

We restrict our discussion of the eyes to two phenomena, papilledema and subhyaloid hemorrhage. Increased intracranial pressure occurs when the contents of the cranium exceed the capacity of the intracranial physiologic mechanisms and anatomy to accommodate additional space-occupying processes. The major accommodating factors are the cerebrospinal fluid space and its ability to be drained by the venous sinuses, the venous space and its collapsibility, the ability of sutures to spread in infants and toddlers, the ability of brain tissue to be compressed and lose substance, the ability of the foramen magnum (and to a lesser degree other foramina) to transmit pressure to the extracranial spaces, and, finally, the possibility of decreased production of cerebrospinal fluid from the choroid plexi when intracranial pressure rises to high levels. The major causes of increased intracranial pressure are cerebral edema, acute hydrocephalus (blockage of cerebrospinal fluid [CSF] absorption, relative or absolute), mass lesions (e.g., neoplasm, abscess, hemorrhage), and venous occlusion (e.g., sagittal or lateral sinus thrombosis).

Papilledema or edema of the optic disk usually indicates increased intracranial pressure. When it is fully developed, recognition is not difficult; swollen and elevated disk edges, engorged and pulseless veins, and increased vascularity of the disk margins are the obvious signs. With further development, hemorrhage (both superficial and deep) and exudates appear. If the process is chronic, filmy white strands of glia proliferate in and around the disk. It is at this late stage that the patient may complain of episodic obscured vision. This precedes final occlusion of the retinal arterial supply and infarction of the retina with permanent blindness. Recognition of this possibility in its early symptomatic stages demands appropriate intracranial medical and/or surgical decompression procedures.

The early, subtle signs of intracranial hypertension should be learned. Prior to well-established and easily recognizable papilledema, the usually present (approximately 75% of population) or easily elicited (approximately 25%) *venous pulsations* disappear. These are best seen in the normal fundus where the veins disappear into the substance of the disk. They reflect the arterial pulse pressure superimposed on a baseline intraocular pressure; the veins partially collapse during systole and expand during diastole. If the pulsations are not spontaneously present, a minimal amount of pressure on the globe brings them out in almost all persons

who do not have increased intracranial pressure (less than 200 mm of CSF). The minimal compression partially collapses the veins and allows them to expand during diastole. If intracranial pressure is 200 mm of CSF or greater, venous pulsation usually is not present but sometimes can be elicited by firm pressure on the globe. The mechanism for loss of venous pulsations is presumed to be an increase in venous backpressure subsequent to intracranial hypertension. However, venous pulsations may be lost or suppressed with a depression of intraocular pressure (e.g., immediately following removal of the lens for cataracts), and if the low global pressure is sustained, full-fledged papilledema occasionally develops. Therefore, it can be assumed that papilledema is a function of the ratio of intracranial pressure to intraocular pressure; elevation of the former or depression of the latter is adequate to elicit edema of the disk. For practical purposes, papilledema is almost always the result of increased intracranial pressure. Increased intraocular pressure (glaucoma) should delay the appearance of papilledema, and this is so; it can be the source of some diagnostic confusion.

The retina is very sensitive to mechanical pressure. You may demonstrate this by pressing very lightly on the lateral side of one of your eyes. The depolarization block caused by minimal compression of the retina creates a blind spot (scotoma) in the contralateral field (i.e., next to your nose). In like manner, early and poorly visible swelling of the disk margin depolarizes and blocks the proximate retina and enlarges the physiologic blind spot. The blind spot represents the retina-deficient optic disk and is routinely plotted and of fairly uniform size when formal visual fields are studied with a tangent screen or perimeter (see chap. 3).

Papillitis or inflammatory edema of the disk is not easily differentiated from papilledema by fundoscopy. Indeed, in most cases they are identical. Papillitis is most often caused by demyelinating processes in young and middle-aged persons and by optic nerve arterial involvement in older individuals. It is not associated with increased intracranial pressure or decreased orbital pressure. As opposed to papilledema, however, it is almost always unilateral. The visual field loss associated with papillitis is almost invariably central, because the macular or cone vision fibers are primarily affected. A central scotoma (blind area) is present and thus visual acuity is severely and uncorrectably limited (see chap. 3). Additionally, because of the disk margin edema, the blind spot is enlarged. No loss of visual acuity occurs with papilledema until quite late when the arterial supply is compromised by compression. This visual loss usually starts at the periphery, with central vision preserved until late.

Subhyaloid hemorrhage is a collection of extravasated blood just beneath the inner limiting membrane of the retina (Fig 1–3). Most retinal hemorrhages occur in the vascular layer of the retina in or deep to the nerve fiber layer. If the hemorrhage is large, it may tear through the nerve fiber layer but then is stopped from diffusing into the vitreous by the inner limiting membrane; or if a hemorrhage is from one of the major retinal veins that lie superficial to the nerve fiber layer, it also pools directly beneath the inner limiting membrane. In persons with long-standing diabetes mellitus or systemic hypertension, these blot-like hemorrhages may be present but are usually associated with other abnormalities of the retina, including hemorrhages of the nerve fiber layer (flame-shaped or striated), narrowing and atherosclerotic distortion of the arteries, exudates, capillary aneurysms, and vascular proliferation (neovascularization). With an *acute, catastrophic rise in intracranial pressure*, almost invariably caused by intracranial arterial hemorrhage (subarachnoid or intracerebral), or head trauma with hemorrhage and brain contusion or laceration, subhyaloid hemorrhages are frequently observed close to the disk margins. They appear almost immediately and frequently on