



FIGURE 104-1 The nervous system can be conceptually reduced to a series of higher-order inputs that converge on final common pathways. For example, upper motor neurons converge on lower motor neurons, whose axons form the final common pathway to an effector muscle.

or corticospinal pathways in the brain stem and spinal cord); dystonia, rigidity, tremor, and tic (i.e., basal ganglia or extrapyramidal systems); and ataxia and dysmetria (i.e., cerebellum). An exception is hypotonia, which is caused by cerebellar disease.

TECHNOLOGIC ASSESSMENT

Laboratory investigations and special testing should be used to confirm a clinical suggestion and to finalize the diagnosis. Testing should be selectively performed because of expense, risk, and discomfort to the patient. Frequently helpful tests are discussed in subsequent sections. Diagnostic tests should never be ordered without a specific differential diagnosis firmly in mind. Many neurodiagnostic tests disclose incidental abnormalities unrelated to a patient's symptomatic disease process.

Lumbar Puncture

Investigation of the cerebrospinal fluid (CSF) is indicated in a small number of specific circumstances, usually meningitis and encephalitis (Table 104-6). When taken, a CSF specimen should be routinely sent for laboratory testing to determine cell and differential counts, protein and glucose levels, and bacterial cultures. The CSF should also be examined for its color and clarity. Cloudy or discolored CSF should be centrifuged and examined for xanthochromia in comparison with water. Additional, special studies may be obtained as appropriate, including Gram stain; fungal, viral, and tuberculous cultures; cryptococcal and other antigens; tests for syphilis; Lyme titers; malignant cytologic patterns; paraneoplastic and other specific protein antibodies; and oligoclonal bands. Polymerase chain reaction for specific viruses may also be appropriate. Assessment of specific CSF proteins such as tau, phosphorylated tau, and amyloid- β in patients at risk for dementia is considered more useful in research than clinical settings. The 14-3-3 protein may be found in patients with rapid-onset dementia.

Recording the opening and closing pressures is important. Tissue infection in the region of the puncture site is an absolute contraindication to lumbar puncture. Relative contraindications

TABLE 104-6 INDICATIONS FOR LUMBAR PUNCTURE

URGENT (DO NOT WAIT FOR BRAIN IMAGING)

Acute central nervous system infection in the absence of focal neurologic signs

LESS URGENT (WAIT FOR BRAIN IMAGING)

Vasculitis, subarachnoid hemorrhage, or cryptic process
Increased intracranial pressure in the absence of mass lesion on magnetic resonance imaging or computed tomography
Intrathecal therapy for fungal or carcinomatous meningitis
Symptomatic treatment for headache from idiopathic intracranial hypertension or subarachnoid hemorrhage

include known or probable intracranial or spinal mass lesion, increased intracranial pressure as a result of mass lesions, coagulopathy caused by thrombocytopenia (usually correctable), anti-coagulant therapy, and bleeding disorders.

Rare but severe complications of lumbar puncture include transtentorial or foramen magnum herniation, spinal epidural hematoma, spinal abscess, herniated or infected disk, meningitis, and adverse reaction to a local anesthetic agent. More common and relatively benign complications include headache and backache.

Tissue Biopsies

In selected specialty centers, a diagnostic biopsy is performed on various tissues, including brain, peripheral nerve (see Chapter 121), muscle (see Chapter 121), and skin. Occasionally, biopsy provides the only means of arriving at a definitive diagnosis.

Electrophysiologic Studies

Electrophysiologic studies include electroencephalography, electromyography, nerve conduction studies, and evoked potentials. These studies are helpful in situations in which the patient cannot be examined or interviewed adequately.

Electroencephalography is most often used to investigate seizures (see Chapter 118). It can document encephalopathy, in which case the background electrical activity of the brain is slowed, and it is also used in the evaluation of brain death.

Electromyography is useful in the differential diagnosis of muscle disease, neuromuscular junction disease, peripheral nerve disease, and anterior horn cell disease (see Chapter 121). Nerve conduction studies (see Chapters 122 and 123) may show decreased amplitude (characteristic of axonal neuropathy) or decreased velocity (characteristic of demyelinating neuropathy).

Visual-evoked potential studies are commonly used in the evaluation of possible multiple sclerosis (see Chapter 120). Asymmetrical slowing of the cortical response to visual pattern stimulation suggests demyelination in the optic nerve or central optic pathways. Brain stem auditory-evoked potential studies are useful in the diagnosis of diseases affecting cranial nerve VIII or its central projections. Lesions at the cerebellopontine angle and the brain stem cause abnormal delay in conduction. Brain stem auditory-evoked potentials are helpful in the diagnosis of deafness in infants. Somatosensory-evoked potentials are used to identify a slowing of central sensory conduction that results from demyelinating disease, compression, or metabolic derangements.