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Neurologic disorders

Introduction

The neurologic system, the body's communications network, coordinates and organizes the functions of all body systems. This intricate network has three main divisions:

- *central nervous system (CNS):* the control center, made up of the brain, the brain stem, and the spinal cord
- *peripheral nervous system:* motor and sensory nerves that connect the CNS to remote body parts and relay and receive messages from them
- *autonomic nervous system (part of the peripheral nervous system):* regulates involuntary functioning of the internal organs and vascular system.

Fundamental unit

The fundamental unit of the nervous system is the neuron, a highly specialized conductor cell that receives and transmits electrochemical nerve impulses. Its structure contains delicate, threadlike nerve fibers that extend from the central cell body and transmit signals: *axons,* which carry impulses *away* from the cell body, and *dendrites,* which carry impulses *to* it. Most neurons have multiple dendrites but only one axon. *Sensory (afferent) neurons* transmit impulses from special receptors to the spinal cord or the brain, *motor (efferent) neurons* transmit impulses from the CNS to regulate activity of muscles or glands, and *interneurons (connecting or association neurons)* shuttle signals

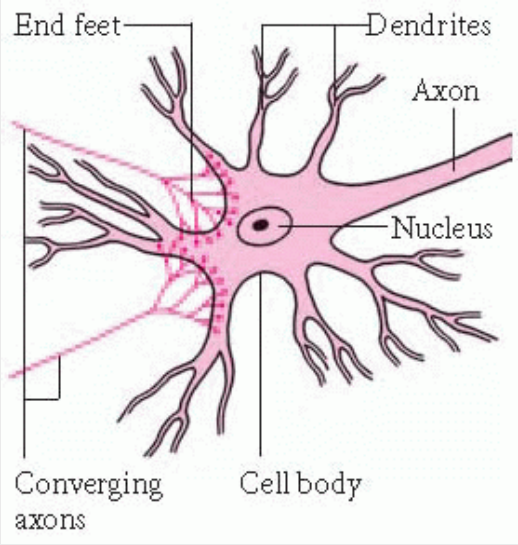
through complex pathways between sensory and motor neurons. Interneurons account for 99% of all the neurons in the nervous system and include most of the neurons in the brain. (See *Structure of the neuron*.)

Intricate control system

This intricate network of interlocking receptors and transmitters forms, together with the brain and spinal cord, a dynamic control system—a living computer—that controls and regulates every mental and physical function. From birth to death, this astonishing system efficiently organizes the body's affairs—controlling the smallest action, thought, or feeling; monitoring communication and instinct for survival; and allowing introspection, wonder, abstract thought, and awareness of one's own intelligence. The brain, the primary center of the CNS, is the large soft mass of nervous tissue housed in the cranium and protected and supported by the meninges and skull bones.

STRUCTURE OF THE NEURON

The basic structure of the neuron is composed of the cell body, axon, and dendrites, as depicted below.



The fragile brain, brain stem, and spinal cord are protected by bone (the skull and vertebrae), cushioning *cerebrospinal fluid (CSF),* and three protective membranes, called *meninges:*

- The *dura mater,* or outer sheath, is made of tough white fibrous tissue.
- The *arachnoid membrane,* the middle layer, is delicate and lacelike.
- The *pia mater,* the inner meningeal layer, is made of fine blood vessels held together by connective tissue. It's thin and transparent and clings to the brain and spinal cord surfaces, carrying branches of the cerebral arteries deep into the brain's fissures and sulci.

Between the dura mater and the arachnoid membrane is the *subdural space;* between the pia mater and the arachnoid membrane is the *subarachnoid space*. The subarachnoid space and the brain's four ventricles contain *CSF,* a clear liquid containing

water and traces of organic materials (especially protein), glucose, and minerals.

CSF is formed from blood in capillary networks called *choroid plexus,* which are located primarily in the brain's lateral ventricles. This fluid is eventually reabsorbed into the venous blood through the *arachnoid villi,* in dural sinuses on the brain's surface.

The *cerebrum,* the largest portion of the brain, is the nerve center that controls sensory and motor activities and intelligence. The outer layer of the cerebrum, the *cerebral cortex*, consists of neuron cell bodies *(gray matter);* the inner layers consist of axons *(white matter)* and basal ganglia, which control motor coordination and steadiness. The cerebral surface is deeply convoluted, furrowed with elevations *(gyri)* and depressions *(sulci)*. A *longitudinal fissure* divides the cerebrum into two hemispheres connected by a wide band of nerve fibers called the *corpus callosum,* through which the hemispheres share information. The hemispheres don't share equally; one always dominates, giving one side control over the other. Because motor impulses descending from the brain through the pyramidal tract cross in the medulla, the right hemisphere controls the left side of the body; the left hemisphere, the right side of the body. Several fissures divide the cerebrum into lobes, each of which is associated with specific functions. (See *A look at the lobes*.)

The *thalamus,* a relay center below the corpus callosum, further organizes cerebral function by transmitting impulses to and from

appropriate areas of the cerebrum. In addition to its primary relay function, it's responsible for primitive emotional responses, such as fear, and for distinguishing pleasant stimuli from unpleasant ones.

The *hypothalamus,* which lies beneath the thalamus, is an autonomic center that has connections with the brain, spinal cord, autonomic nervous system, and pituitary gland. It regulates temperature control, appetite, blood pressure, breathing, sleep patterns, and peripheral nerve discharges that occur with behavioral and emotional expression. It also has partial control of pituitary gland secretion and stress reaction.

The base of the brain

Beneath the cerebrum, at the base of the brain, is the *cerebellum*. It's responsible for smooth muscle movements, coordinating sensory impulses with muscle activity, and maintaining muscle tone and equilibrium.

The brain stem houses cell bodies for most of the cranial nerves and includes the *midbrain,* the *pons,* and the *medulla oblongata*. With the thalamus and the hypothalamus, it makes up a nerve network called the *reticular formation,* which acts as an arousal mechanism and controls wakefulness. It also relays nerve impulses between the spinal cord and other parts of the brain. The midbrain is the reflex center for the third and fourth cranial nerves and mediates pupillary reflexes and eye movements. The pons helps regulate respirations; it's also the reflex center for the fifth through eighth cranial nerves and mediates chewing, taste, saliva secretion, hearing, and equilibrium. The medulla oblongata affects cardiac, respiratory, and vasomotor functions.

Bloodline to the brain

Four major arteries—two vertebral and two carotid—supply the brain with oxygenated blood. These arteries originate in or near the aortic arch. The two vertebral arteries (branches of the subclavian) converge to become the basilar artery, which supplies the posterior brain. The common carotids branch into the two internal carotids, which divide further to supply the anterior brain and the middle brain. These arteries interconnect through the *Circle of Willis,* at the base of the brain. This anastomosis usually ensures continual circulation to the brain.

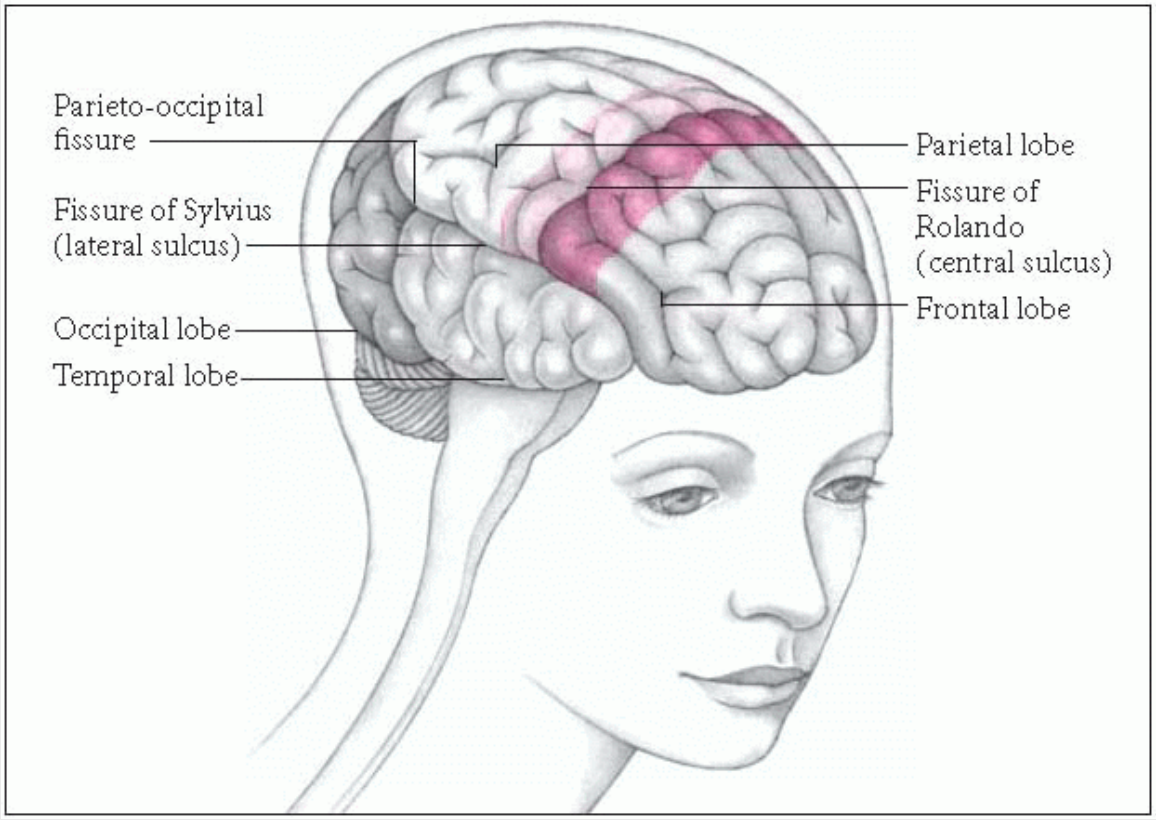
The spinal cord: Conductor pathway

Extending downward from the brain through the vertebrae, to the level of approximately the second lumbar vertebra, is the spinal cord, a two-way conductor pathway between the brain stem and the peripheral nervous system. The spinal cord is also the reflex center for activities that

don't require brain control, such as deep tendon reflexes, the jerking reaction elicited by tapping with a reflex hammer.

A LOOK AT THE LOBES

Several fissures divide the cerebrum into hemispheres and lobes; each lobe has a specific function. The *fissure of Sylvius* (lateral sulcus) separates the temporal lobe from the frontal and parietal lobes. The *fissure of Rolando* (central sulcus) separates the frontal lobes from the parietal lobe. The *parieto-occipital fissure* separates the occipital lobe from the two parietal lobes.



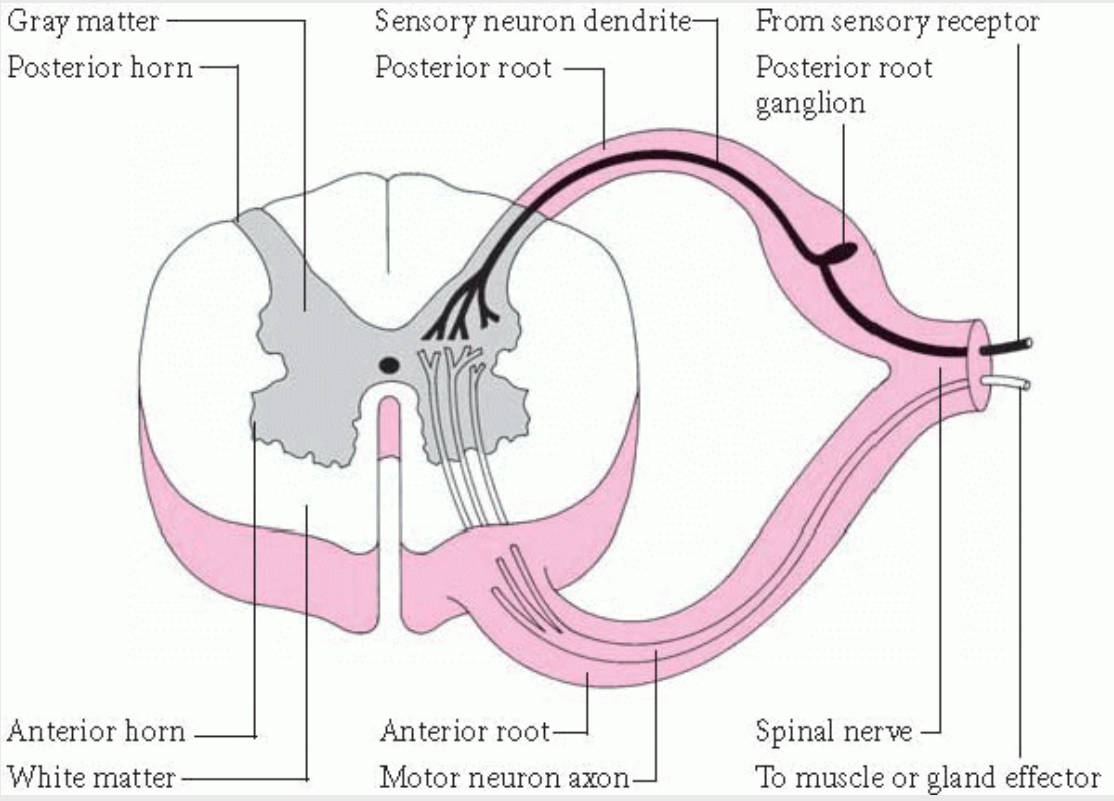
- The *frontal lobe* controls voluntary muscle movements and contains motor areas (including the motor area for speech, or Broca's area). It's the center for personality, behavioral, and intellectual functions, such as judgment, memory, and problem solving; for autonomic functions; and for cardiac and emotional responses.
- The *temporal lobe* is the center for taste, hearing, and smell, and in the brain's dominant hemisphere, it interprets spoken language.
- The *parietal lobe* coordinates and interprets sensory information from the opposite side of the body.
- The *occipital lobe* interprets visual stimuli.

A cross section of the spinal cord shows an internal H-shaped mass of gray matter divided into horns, which consist primarily of neuron cell bodies. (See *Cross section of the spinal cord,* page 168.) Cell bodies in

the posterior, or dorsal, horn primarily relay sensations; those in the anterior, or ventral, horn are needed for voluntary or reflex motor activity. The white matter surrounding the outer part of these horns consists of myelinated nerve fibers grouped functionally in vertical columns, called *tracts*. The sensory, or ascending, tracts carry sensory impulses up the spinal cord to the brain; the motor, or descending, tracts carry motor impulses down the spinal cord. The brain's motor impulses reach a descending tract and continue through the peripheral nervous system via upper motor neurons. These neurons originate in the brain and form two major systems:

CROSS SECTION OF THE SPINAL CORD

The cross section of the spinal cord below shows the anterior and posterior segments.



- The *pyramidal system* (corticospinal tract) is responsible for fine, skilled movements of skeletal muscle. An impulse in this system originates in the frontal lobe's motor cortex and travels downward to

the pyramids of the medulla, where it crosses to the opposite side of the spinal cord.

- The *extrapyramidal system* (extracorticospinal tract) controls gross motor movements. An impulse traveling in this system originates in the frontal lobe's motor cortex and is mediated by basal ganglia, the thalamus, cerebellum, and reticular formation before descending to the spinal cord.

Outlying areas

Messages transmitted through the spinal cord reach outlying areas through the peripheral nervous system, which originates in 31 pairs of segmentally arranged spinal nerves attached to the spinal cord. Spinal nerves are numbered according to their point of origin in the cord:

- 8 cervical: C1 to C8
- 12 thoracic: T1 to T12
- 5 lumbar: L1 to L5
- 5 sacral: S1 to S5
- 1 coccygeal.

On the cross section of the spinal cord, you'll see that these spinal nerves are attached to the spinal cord by two roots:

- The *anterior,* or *ventral*, root consists of motor fibers that relay impulses from the cord to glands and muscles.
- The *posterior,* or *dorsal,* root consists of sensory fibers that relay sensory information from receptors to the cord. The posterior root has an enlarged area—the posterior root ganglion—which is made up of sensory neuron cell bodies.

After leaving the vertebral column, each spinal nerve separates into *rami* (branches), distributed peripherally, with extensive but organized overlapping. This overlapping reduces the chance of lost sensory or motor function from interruption of a single spinal nerve.

Two functional systems

- The *somatic (voluntary) nervous system* is activated by will but can also function independently. It's responsible for all conscious and higher mental processes and for subconscious and reflex actions such as shivering.
- *The autonomic (involuntary) nervous system* regulates unconscious processes to control involuntary body functions, such as digestion, respiration, and cardiovascular function. It's usually divided into two competing systems: The sympathetic nervous system controls energy expenditure, especially in stressful situations, by releasing adrenergic catecholamines. The parasympathetic nervous system helps conserve energy by releasing the cholinergic neurohormone acetylcholine. These systems balance each other to support homeostasis under normal conditions.

Assessing neurologic function

A complete neurologic assessment helps confirm the diagnosis when a neurologic disorder is suspected. It establishes a clinical baseline and can offer lifesaving clues to rapid deterioration. Neurologic assessment includes:

- *Patient history:* In addition to the usual information, try to elicit the patient's and his family's perception of the disorder. Use the patient interview to make observations that help evaluate mental status and behavior.
- *Physical examination:* Pay particular attention to obvious abnormalities that may signal serious neurologic problems; for example, fluid draining from the nose or ears. Check for these significant symptoms:

  – headaches, especially if they're more severe in the morning, wake the patient, or the pain is unusually intense

  – change in visual acuity, especially sudden change

  – numbness or tingling in one or more extremities

  – clumsiness or complete loss of function in an extremity

- mood swings or personality changes
- any change in seizures.

- *Neurologic examination:* Determine cerebral, cerebellar, motor, sensory, and cranial nerve function.

Obviously, there isn't always time for a complete neurologic examination during bedside assessment. It's therefore necessary to select priorities; for example, typical bedside assessment focuses on level of consciousness, pupillary response, motor function, reflexes, sensory functions, and vital signs. However, when time permits, a complete neurologic examination can provide valuable information regarding total neurologic function.

Assessing mental status, intellect, and behavior

Mental status and behavior are good indicators of cerebral function, and they're easy to assess. Note the patient's appearance, mannerisms, posture, facial expression, grooming, and tone of voice. Check for orientation to time, place, and person and for memory of recent and past events. To test intellect, ask the patient to count backward from 100 by 7s, to read aloud, or to interpret a common proverb, and see how well he understands and follows commands. To assesses executive function, ask the patient to follow a series of commands. If you make such checks frequently, vary the questions to avoid a programmed response.

Assessing level of consciousness

Level of consciousness (LOC) is a valuable indicator of neurologic function. It can vary from alertness (response to verbal stimulus) to coma (failure to respond even to painful stimulus). Document the patient's exact response to the stimulus; for example, write, “Patient pulled away in response to nail bed pressure,” rather than a simple adjective like “stuporous.”

The Glasgow Coma Scale (GCS), which assesses eye opening as well as verbal and motor responses, provides a quick, standardized account of neurologic status. In this test, each response receives a numerical value. (See *Glasgow Coma Scale,* page 170.) For instance, if the patient readily

responds verbally and is oriented to time, place, and person, he scores a 5; if he's completely unable to respond verbally, he scores a 1. If the patient is intubated or has a tracheostomy, assess and score accordingly, such as 5/T ('T' meaning tracheostomy). A score of 15 for all three parts is normal; 7 or less indicates coma; 3—the lowest score possible —generally (but not always) indicates brain death. Although the GCS is useful, it isn't a substitute for a complete neurologic assessment.

GLASGOW COMA SCALE

To quickly assess a patient's level of consciousness and to uncover baseline changes, use the Glasgow Coma Scale. This assessment tool grades consciousness in relation to eye opening and motor and verbal responses. A decreased reaction score in one or more categories warns of an impending neurologic crisis. A patient scoring 7 or less is comatose and probably has severe neurologic damage.

Test	Patient's reaction	Score
Best eye opening response	Open spontaneously	4
	Open to verbal command	3
	Open to pain	2
	No response	1
Best motor response	Obeys verbal command	6
	Localizes painful stimuli	5
	Flexion-withdrawal	4
	Flexion-abnormal (decorticate rigidity)	3
	Extension (decerebrate rigidity)	2

	No response	1
Best verbal response	Oriented and converses	5
	Disoriented and converses	4
	Inappropriate words	3
	Incomprehensible sounds	2
	No response	1
Total		**3 to 15**

Assessing motor function

The inability to perform the following simple tests, or the presence of tics, tremors, or other abnormalities during such testing, suggests cerebellar dysfunction.

- Ask the patient to touch his nose with each index finger, alternating hands. Repeat this test with his eyes closed.
- Instruct the patient to tap the index finger and thumb of each hand together rapidly.
- Have the patient draw a figure eight in the air with his foot.
- To test tandem walk, ask the patient to walk heel to toe in a straight line.
- To test balance, perform the Romberg test: Ask the patient to stand with feet together, eyes closed, and arms outstretched without losing balance.

Motor function is a good indicator of LOC and can also point to central or peripheral nervous system damage. During all tests of motor function, watch for differences between right and left side functions.

- To check gait, ask the patient to walk while you observe posture, balance, and coordination of leg movement and arm swing.

- To check muscle tone, palpate muscles at rest and in response to passive flexion. Look for flaccidity, spasticity, and rigidity. Measure muscle size, and look for involuntary movements, such as rapid jerks, tremors, or contractions.
- To evaluate muscle strength, have the patient grip your hands and squeeze. Then ask him to push against your palm with his foot. Compare muscle strength on each side, using a 5-point scale (5 is normal strength, 0 is complete paralysis). Also test the patient's ability to extend and flex the neck, elbows, wrists, fingers, toes, hips, and knees; to extend the spine; to contract and relax the abdominal muscles; and to rotate the shoulders.
- Rate reflexes on a 4-point scale (4 is clonus, 0 is absent reflex). Before testing reflexes, make sure that the patient is comfortable and relaxed. Then, to test superficial reflexes, stroke the skin of the abdominal, gluteal, plantar, and scrotal regions with a moderately sharp object that won't puncture the skin. (Don't use the same pin to test another patient.) A normal response to this stimulus is flexion. To test deep reflexes, use a reflex hammer to briskly tap the biceps, the triceps, and the brachioradialis, patellar, and Achilles tendon regions. A normal response is rapid extension and contraction.

Assessing sensory function

Impaired or absent sensation in the trunk or extremities can point to brain, spinal cord, or peripheral nerve damage. Determining the extent of sensory dysfunction is important because it helps locate neurologic damage. For instance, localized dysfunction indicates local peripheral nerve damage, dysfunction over a single dermatome (an area served by 1 of the 31 pairs of spinal nerves) indicates damage to the nerve's dorsal root, and dysfunction extending over more than one dermatome suggests brain or spinal cord damage.

In assessing sensory function, always test both sides of symmetrical areas—for instance, both arms, not just one. Reassure the patient that the test won't be painful.

- *Superficial pain perception:* Lightly press the point of an open safety pin against the patient's skin. Don't press hard enough to scratch the skin. Discard pin after use.

- *Thermal sensitivity:* The patient tells what he feels when you place a test tube filled with hot water and one filled with cold water against his skin.
- *Tactile sensitivity:* Ask the patient to close his eyes and tell you what he feels when touched lightly on hands, wrists, arms, thighs, lower legs, feet, and trunk with a wisp of cotton.
- *Sensitivity to vibration:* Place the base of a vibrating tuning fork against the patient's wrists, elbows, knees, or other bony prominences. Hold it in place, and ask the patient to tell you when it stops vibrating.
- *Position sense:* Hold the lateral medial portion of the patient's fingers and toes and move them up, down, and to the side. Ask the patient to tell you the direction of movement.
- *Discriminatory sensation:* Ask the patient to close his eyes and identify familiar textures (velvet or burlap) or objects placed in his hand or numbers and letters traced on his palm.
- *Two-point discrimination:* Using calipers or other sharp objects, touch the patient in two different places simultaneously. Ask if he can feel one or two points. Record how many millimeters of separation are required for the patient to feel two points.

Assessing cranial nerve function

By using the simple tests that follow, you can reliably localize cranial nerve dysfunction.

- *Olfactory nerve (I):* Have the patient close his eyes and, using each nostril separately, try to identify common nonirritating smells, such as cinnamon, coffee, or peppermint.
- *Optic nerve (II):* Examine the patient's eyes with an ophthalmoscope, and have him read a Snellen eye chart or a newspaper. To test peripheral vision, ask him to cover one eye and fix his other eye on a point directly in front of him. Then, ask if he can see you wiggle your finger in the

  four quadrants; you'd expect him to see your finger in all four.

- *Oculomotor nerve (III):* Compare the size and shape of the patient's pupils and the equality of pupillary response to a small light in a darkened room. Shine the light from a lateral position, not directly in front of the patient's eyes.
- *Trochlear nerve (IV) and abducens nerve (VI):* To assess for conjugate and lateral eye movement, ask the patient to follow your finger with his eyes as you slowly move it from his far left to his far right.
- *Trigeminal nerve (V):* To test all three portions of this cranial nerve, test facial sensation by stroking the patient's jaws, cheeks, and forehead with a cotton swab, the point of a pin, or test tubes filled with hot or cold water. Because testing for a blink reflex is irritating to the patient, it's not commonly done. If you must test for this response (it may be decreased in patients who wear contact lenses), touch the cornea lightly with a wisp of cotton or tissue, and avoid repeating the test, if possible. To test for jaw jerk, ask the patient to hold his mouth slightly open; then tap the middle of his chin lightly with a reflex hammer. The jaw should jerk closed.
- *Facial nerve (VII):* To test upper and lower facial motor function, ask the patient to raise his eyebrows, close his eyes, wrinkle his forehead, and show his teeth. To test sense of taste, ask him to identify the taste of salty, sour, sweet, and bitter substances, which you have placed on his tongue.
- *Acoustic nerve (VIII):* Ask the patient to identify common sounds such as a ticking clock. With a tuning fork, test for air and bone conduction.
- *Glossopharyngeal nerve (IX):* To test gag reflex, touch a tongue blade to each side of the patient's pharynx.
- *Vagus nerve (X):* Observe ability to swallow, and watch for symmetrical movements of soft palate when the patient says, “Ah.”
- *Spinal accessory nerve (XI):* To test shoulder muscle strength, palpate the patient's shoulders, and ask him to shrug against a resistance.
- *Hypoglossal nerve (XII):* To test tongue movement, ask the patient to stick out his tongue. Inspect it for tremor, atrophy, or lateral deviation. To test for strength, ask the patient to move his tongue from side to side while you hold a tongue blade against it.

Testing for a firm diagnosis

A firm diagnosis of many neurologic disorders usually requires a wide range of diagnostic tests—both noninvasive and invasive. Noninvasive tests are done first and may include the following:

- Skull X-ray identifies skull malformations, fractures, erosion, or thickening. Changes in landmarks may indicate a space-occupying lesion.
- Computed tomography (CT) scan produces three-dimensional images that can identify hemorrhage, intracranial tumors, malformation, and cerebral atrophy, edema, calcification, and infarction. If a contrast medium is used, the procedure is invasive.
- Magnetic resonance imaging (MRI) views the CNS in greater detail than a CT scan and is the procedure of choice for detecting multiple sclerosis; intraluminal clots and blood flow in arteriovenous malformations and aneurysms; brain stem, posterior fossa, and spinal cord lesions; early cerebral infarction; and brain tumors. A noniodinated contrast medium may be used to enhance lesions. Advances in MRI allow visualization of cerebral arteries and venous sinuses without administration of a contrast medium.
- EEG detects abnormal electrical activity in the brain (for example, from a seizure, metabolic disorder, or drug overdose).
- Ultrasonography detects carotid lesions or changes in carotid blood flow and velocity. High-frequency sound waves reflect back the velocity of blood flow, which is then reported as a graphic recording of a waveform.
- Evoked potentials evaluate the visual, auditory, and somatosensory nerve pathways by measuring the brain's electrical response to stimulation of the sensory organs or peripheral nerves.

Invasive tests may include the following:

- In lumbar puncture, a needle is inserted into the subarachnoid space of the spinal cord, usually between L3 and L4 (or L4 and L5). This allows aspiration of CSF for

analysis to detect infection or hemorrhage; to determine cell count and glucose, protein, and globulin levels; and to measure CSF pressure. Lumbar puncture is usually contraindicated in hydrocephalus and in increased intracranial pressure (ICP) because a quick pressure reduction may cause brain herniation. (See *What happens in increased ICP,* page 174.)

- Myelography follows a lumbar puncture and CSF removal. In this procedure, a radiologic dye is instilled and X-rays show spinal abnormalities and determine spinal cord compression related to back pain or extremity weakness.
- In cerebral arteriography, also known as *angiography,* a catheter is inserted into an artery—usually the femoral artery—and is threaded up to the carotid artery. Then a radiopaque dye is injected, allowing X-ray visualization of the cerebral vasculature. Sometimes the catheter is threaded directly into the brachial or carotid artery. This test can show cerebrovascular abnormalities and spasms plus arterial changes due to a tumor, arteriosclerosis, hemorrhage, an aneurysm, or blockage. A patient undergoing this procedure is at risk for a stroke and for increased ICP.
- Digital subtraction angiography visualizes cerebral vessels using contrast medium administered I.V., after which computer-assisted precontrast and postcontrast images are compared. The first image is "subtracted" from the second, which highlights the cerebral vessels.
- Brain scan measures gamma rays produced by a radioisotope injected I.V. Uptake and distribution of the isotope in the brain highlights intracranial masses, vascular lesions, and other problems.
- ICP monitoring can be a direct, invasive method of identifying trends in ICP. A subarachnoid screw and an intraventricular catheter convert CSF pressure readings into waveforms that are displayed digitally on an oscilloscope monitor. Another method uses a fiber-optic catheter inserted in the subdural space; with this indirect method, pressure changes are reported digitally or in waveform.
- Electromyography detects lower motor neuron disorders, neuromuscular disorders, and nerve damage. A needle inserted into

selected muscles at rest and during voluntary contraction picks up nerve impulses and measures nerve conduction time.

CONGENITAL ANOMALIES

Cerebral palsy

The most common cause of crippling in children, cerebral palsy (CP) is a group of neuromuscular disorders resulting from prenatal, perinatal, or postnatal CNS damage. Although nonprogressive, these disorders may become more obvious as an affected infant grows older. Three major types of CP occur—spastic, athetoid, and ataxic—sometimes in mixed forms. Motor impairment may be minimal (sometimes apparent only during physical activities such as running) or severely disabling. Associated defects, such as seizures, speech disorders, and mental retardation, are common. The prognosis varies; in cases of mild impairment, proper treatment may make a near-normal life possible.

Causes and incidence

See *Causes of cerebral palsy*, page 175, for a more detailed description of the causes of CP. Incidence is slightly higher in premature neonates (anoxia plays the greatest role in contributing to CP) and in neonates who are small for their gestational age. CP is slightly more common in males than in females. For every 1,000 births, 2 to 4 neonates are affected.

Spastic cerebral palsy is the most common type of CP, affecting about 50% of CP patients. Athetoid cerebral palsy affects about 20% of CP patients, ataxic cerebral palsy accounts for another 10% of these patients, and the remaining 20% of patients are mixed, with a combination of symptoms.

Complications

- Seizure disorder
- Injuries from falls
- Speech, vision, and hearing problems

- Language and perception deficits
- Respiratory problems (poor swallowing and gag reflexes)
- Dental problems
- Mental retardation

PATHOPHYSIOLOGY

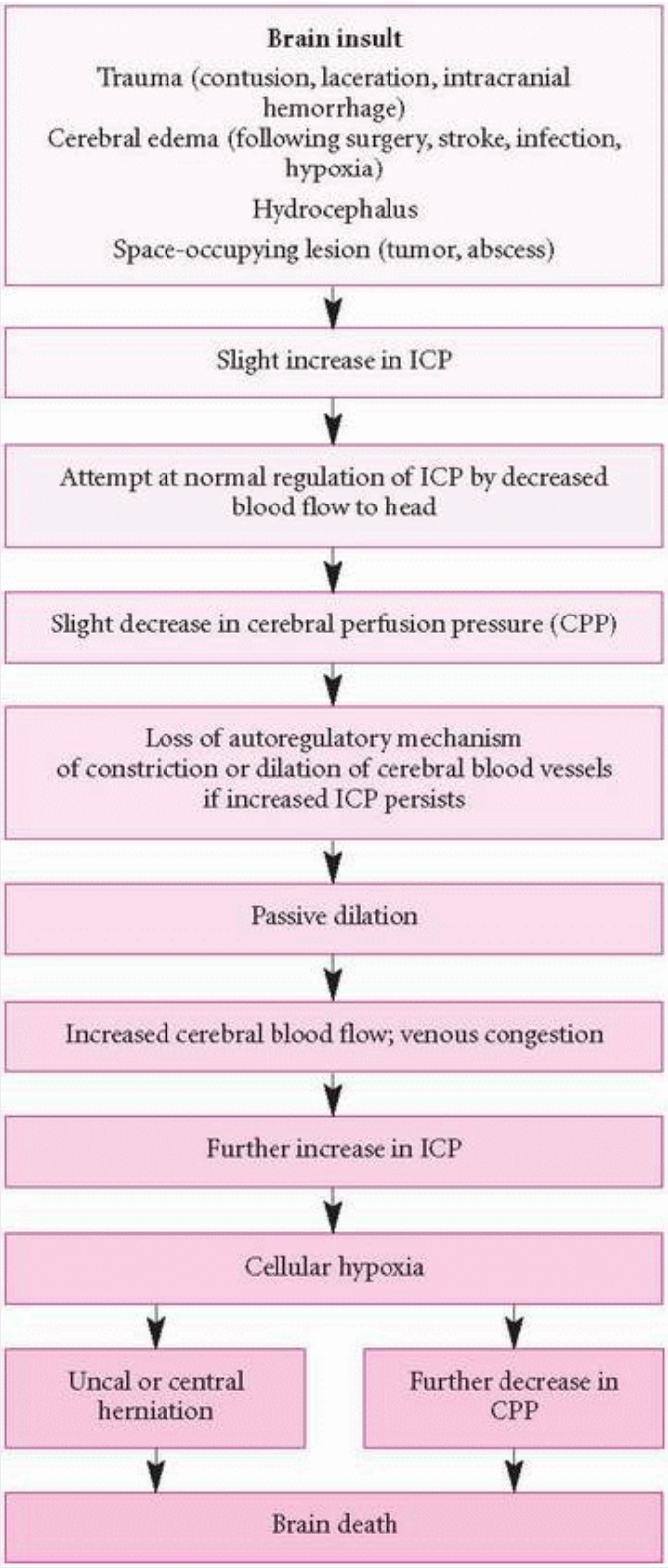
WHAT HAPPENS IN INCREASED ICP

Intracranial pressure (ICP) is the pressure exerted within the intact skull by the intracranial volume—about 10% blood, 10% cerebrospinal fluid (CSF), and 80% brain tissue water. The rigid skull allows very little space for expansion of these substances. When ICP increases to pathologic levels, brain damage can result.

The brain compensates for increases in ICP by regulating the volumes of the three substances in the following ways:

- limiting blood flow to the head
- displacing CSF into the spinal canal
- increasing absorption or decreasing production of CSF—withdrawing water from brain tissue into the blood and excreting it through the kidneys. When compensatory mechanisms become overworked, small changes in volume lead to large changes in pressure.

The chart at right will help you understand the pathophysiology of increased ICP.


Brain insult
Trauma (contusion, laceration, intracranial hemorrhage)
Cerebral edema (following surgery, stroke, infection, hypoxia)
Hydrocephalus
Space-occupying lesion (tumor, abscess)
Slight increase in ICP
Attempt at normal regulation of ICP by decreased blood flow to head
Slight decrease in cerebral perfusion pressure (CPP)
Loss of autoregulatory mechanism of constriction or dilation of cerebral blood vessels if increased ICP persists
Passive dilation
Increased cerebral blood flow; venous congestion
Further increase in ICP
Cellular hypoxia
Uncal or central herniation
Further decrease in CPP
Brain death

Signs and symptoms

Spastic cerebral palsy is characterized by hyperactive deep tendon reflexes, increased stretch reflexes, rapid alternating muscle contraction and relaxation, muscle weakness, underdevelopment of affected limbs, muscle contraction in response to manipulation, and a tendency to contractures. Typically, a child with spastic CP walks on his toes with a scissors gait, crossing one foot in front of the other.

In athetoid cerebral palsy, involuntary movements—grimacing, wormlike writhing, dystonia, and sharp jerks—impair voluntary movement. Usually, these involuntary movements affect the arms more severely than the legs; involuntary facial movements may make speech difficult. These athetoid movements become more severe during stress, decrease with relaxation, and disappear entirely during sleep.

Ataxic cerebral palsy is characterized by disturbed balance, incoordination (especially of the arms), hypoactive reflexes, nystagmus, muscle weakness, tremor, lack of leg movement during infancy, and a widebased gait as the child begins to walk. Ataxia makes sudden or fine movements almost impossible.

Some children with CP display a combination of these clinical features. In most, impaired motor function makes eating (especially swallowing) difficult and retards growth and development. Up to 40% of these children are mentally retarded, about 25% have seizure disorders, and about 80% have impaired speech. Many also have dental abnormalities, vision and hearing defects, and reading disabilities.

Diagnosis

Early diagnosis is essential for effective treatment and requires precise neurologic assessment and careful clinical observation during infancy. Computed tomography scan and magnetic resonance imaging can reveal structural or congenital abnormalities. Suspect CP whenever an infant:

CAUSES OF CEREBRAL PALSY

Conditions that result in cerebral anoxia, hemorrhage, or other damage are probably responsible for cerebral palsy.

- Prenatal conditions that may increase risk of CP: maternal infection (especially rubella), maternal drug ingestion, radiation, anoxia, toxemia, maternal diabetes, abnormal placental attachment, malnutrition, and isoimmunization
- Perinatal and birth difficulties that increase the risk of CP: forceps delivery, breech presentation, placenta previa, abruptio placentae, metabolic or electrolyte disturbances, abnormal maternal vital signs from general or spinal anesthetic, prolapsed cord with delay in delivery of head, premature birth, prolonged or unusually rapid labor, and multiple birth (especially infants born last in a multiple birth)
- Infection or trauma during infancy: poisoning, severe kernicterus resulting from erythroblastosis fetalis, brain infection, head trauma, prolonged anoxia, brain tumor, cerebral circulatory anomalies causing blood vessel rupture, and systemic disease resulting in cerebral thrombosis or embolus

- has difficulty sucking or keeping the nipple or food in his mouth
- seldom moves voluntarily or has arm or leg tremors with voluntary movement
- crosses his legs when lifted from behind rather than pulling them up or “bicycling” like a normal infant
- has legs that are difficult to separate, making diaper changing difficult
- persistently uses only one hand or, as he gets older, uses hands well but not legs.

Infants at particular risk include those with low birth weight, low Apgar scores at 5 minutes, seizures, and metabolic disturbances. However, all infants should have a

screening test for CP as a regular part of their 6-month checkup.

Treatment

CP can't be cured, but proper treatment can help affected children reach their full potential within the limitations set by this disorder. Such treatment requires a comprehensive and cooperative effort involving physicians, nurses, teachers, psychologists, the child's family, and occupational, physical, and speech therapists. Home care is usually possible. Treatment usually includes interventions that encourage optimum development:

- Braces or splints and special appliances, such as adapted eating utensils and a low toilet seat with arms, help these children perform activities independently.
- An artificial urinary sphincter may be indicated for the incontinent child who can use the hand controls.
- Range-of-motion stretches minimize contractures.
- Orthopedic surgery may be indicated to correct contractures. Botulinum toxin has been shown to reduce or delay the need for surgery.
- Phenytoin, phenobarbital, or another anticonvulsant may be used to control seizures.
- Muscle relaxants or neurosurgery may be required to decrease spasticity.

Children with milder forms of CP should attend a regular school; severely afflicted children may need special education.

Special considerations

- A child with CP may be hospitalized for orthopedic surgery or for treatment of other complications.
- Speak slowly and distinctly. Encourage the child to ask for things he wants. Listen patiently and don't rush him.

- Plan a high-calorie diet that's adequate to meet the child's high-energy needs.
- During meals, maintain a quiet, unhurried atmosphere with as few distractions as possible. The child should be encouraged to feed himself and may need special utensils and a chair with a solid footrest. Teach him to place food far back in his mouth to facilitate swallowing.
- Encourage the child to chew food thoroughly, drink through a straw, and suck on lollipops to develop the muscle control needed to minimize drooling.
- Allow the child to wash and dress independently, assisting only as needed. The child may need clothing modifications.
- Give all care in an unhurried manner; otherwise, muscle spasticity may increase.
- Encourage the child and his family to participate in the plan of care so they can continue it at home.
- Care for associated hearing or visual disturbances, as necessary.
- Give frequent mouth care and dental care, as necessary.
- Reduce muscle spasms that increase postoperative pain by moving and turning the child carefully after surgery; provide analgesics as needed.
- After orthopedic surgery, provide cast care. Reposition the child often, check for foul odor, and ventilate under the cast with a cool air blow-dryer. Use a flashlight to check for skin breakdown beneath the cast. Help the child relax, perhaps by giving a warm bath, before reapplying a bivalved cast.

To help the parents:

- Encourage them to set realistic individual goals.
- Assist in planning crafts and other activities.
- Stress the child's need to develop peer relationships; warn the parents against being overprotective.
- Identify and deal with family stress. The parents may feel unreasonable guilt about their child's handicap and may need

psychological counseling.

- Refer the parents to supportive community organizations. For more information, tell them to contact the United Cerebral Palsy Association or their local chapter.

Hydrocephalus

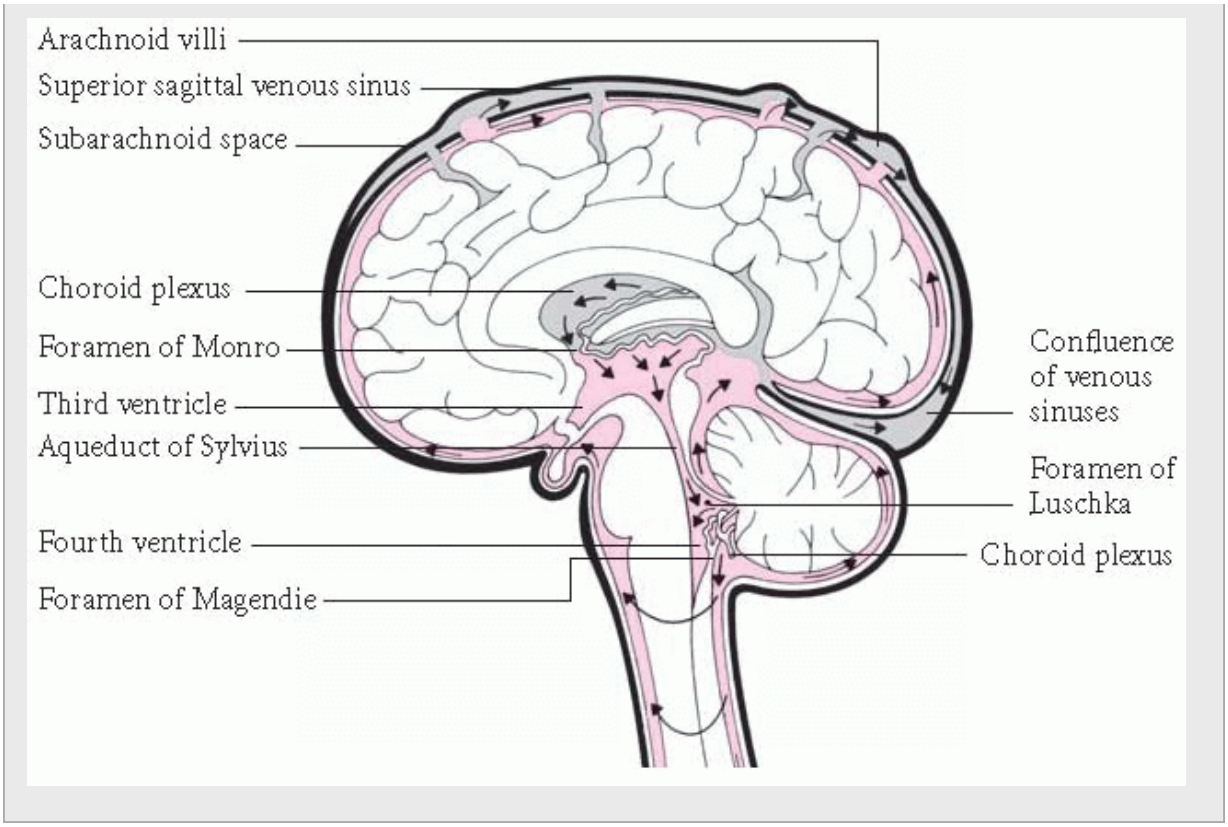
Hydrocephalus is an excessive accumulation of cerebrospinal fluid (CSF) within the ventricular spaces of the brain. In infants, hydrocephalus enlarges the head; in infants and adults, resulting compression can damage brain tissue. With early detection and surgical intervention, the prognosis improves but remains guarded. Even after surgery, such complications as mental retardation, impaired motor function, and vision loss can persist. Without surgery, the prognosis is poor: Mortality may result from increased intracranial pressure (ICP); infants may also die prematurely of infection and malnutrition.

NORMAL CIRCULATION OF CSF

Cerebrospinal fluid (CSF) is produced from blood in a capillary network (choroid plexus) in the brain's lateral ventricles. From the lateral ventricles, CSF flows through the interventricular foramen (foramen of Monro) to the third ventricle. From there, it flows through the aqueduct of Sylvius to the fourth ventricle and through the foramina of Luschka and Magendie to the cisterna of the subarachnoid space.

Then, the fluid passes under the base of the brain, upward over the brain's upper surfaces, and down around the spinal cord. Eventually, CSF reaches the arachnoid villi, where it's reabsorbed into venous blood at the venous sinuses.

Normally, the amount of fluid produced (about 500 ml/day) equals the amount absorbed. The average amount circulated at one time is 150 to 175 ml.



Causes and incidence

Hydrocephalus may result from an obstruction in CSF flow (noncommunicating hydrocephalus) or from faulty absorption of CSF (communicating hydrocephalus). (See *Normal circulation of CSF*.)

In noncommunicating hydrocephalus, the obstruction occurs most frequently between the third and fourth ventricles, at the aqueduct of Sylvius, but it can also occur at the outlets of the fourth ventricle (foramina of Luschka and Magendie) or, rarely, at the foramen of Monro. This obstruction may result from faulty fetal development, infection (syphilis, granulomatous diseases, meningitis), a tumor, cerebral aneurysm, or a blood clot (after intracranial hemorrhage).

In communicating hydrocephalus, faulty absorption of CSF may result from surgery to repair a myelomeningocele, adhesions between meninges at the base of the brain,

or meningeal hemorrhage. Rarely, a tumor in the choroid plexus causes overproduction of CSF, producing hydrocephalus.

SIGNS OF HYDROCEPHALUS

In infants, characteristic changes of hydrocephalus include marked enlargement of the head; distended scalp veins; thin, shiny, and fragile-looking scalp skin; and weak muscles that can't support the head.

Hydrocephalus occurs most commonly in neonates but can also occur in adults as a result of injury or disease. It affects 1 of every 1,000 people.

Complications

- Mental retardation
- Impaired motor function
- Vision loss
- Death

Signs and symptoms

In infants, the unmistakable sign of hydrocephalus is rapidly increasing head circumference, clearly disproportionate to the infant's growth. Other characteristic changes include widening and bulging of the fontanels; distended scalp veins; thin, shiny, and fragile-looking scalp skin; and underdeveloped neck muscles. (See *Signs of hydrocephalus*.) In severe hydrocephalus, the roof of the orbit is depressed, the eyes are

displaced downward, and the sclerae are prominent. Sclera seen above the iris is called the “setting-sun sign.” A high-pitched, shrill cry, abnormal muscle tone of the legs, irritability, anorexia, and projectile vomiting commonly occur. In adults and older children, indicators of hydrocephalus include decreased level of consciousness (LOC), ataxia, incontinence, loss of coordination, and impaired intellect.

Diagnosis

In infants, abnormally large head size for the patient's age strongly suggests hydrocephalus. Measurement of head circumference is a most important diagnostic technique. Skull X-rays show thinning of the skull with separation of sutures and widening of fontanels.

Other diagnostic tests for hydrocephalus, including arteriography, computed tomography scan, and magnetic resonance imaging, can differentiate between hydrocephalus and intracranial lesions and can also demonstrate the Arnold-Chiari deformity, which may occur in an infant with hydrocephalus. (See *Arnold-Chiari syndrome*.)

Treatment

Surgical correction is the only treatment for hydrocephalus. Surgery typically consists of insertion of a ventriculoperitoneal shunt, which transports excess fluid from the lateral ventricle into the peritoneal cavity. A less common procedure is insertion of a ventriculoatrial shunt, which drains fluid from the brain's lateral ventricle into the right atrium of the heart, where the fluid makes its way into the venous circulation.

Complications of surgery include shunt infection, septicemia (after ventriculoatrial shunt), adhesions and paralytic ileus, migration, peritonitis, and intestinal perforation (with peritoneal shunt).

Special considerations

- On initial assessment, obtain a complete history from the patient or his family. Note general behavior, especially irritability, apathy, or decreased LOC. Perform a neurologic assessment. Examine the eyes: pupils should be equal and reactive to light. In adults and older

children, evaluate movements and motor strength in extremities. Watch especially for ataxia, confusion, and incontinence. Ask the patient if he has headaches, and watch for projectile vomiting; both are signs of increased ICP. Also watch for seizures. Note changes in vital signs.

Before surgery to insert a shunt:

- Encourage maternal-infant bonding when possible. When caring for the infant yourself, hold him on your lap for feeding; stroke and cuddle him, and speak soothingly.
- Check fontanels for tension or fullness, and measure and record head circumference. On the patient's chart, draw a picture showing where to measure the head so that other staff members measure it in the same place, or mark the forehead with ink.
- To prevent postfeeding aspiration and hypostatic pneumonia, place the infant on his side and reposition every 2 hours, or prop him up in an infant seat.
- To prevent skin breakdown, make sure his earlobe is flat, and place a sheepskin or rubber foam under his head.
- When turning the infant, move his head, neck, and shoulders with his body to reduce strain on his neck.
- Feed the infant slowly. To lessen strain from the weight of the infant's head on your arm while holding him during feeding, place his head, neck, and shoulders on a pillow.

After surgery:

- Place the infant on the side opposite the operative site with his head level with his body unless the physician's orders specify otherwise.
- Check temperature, pulse rate, blood pressure, and LOC. Also check fontanels for fullness daily. Watch for vomiting, which may be an early sign of increased ICP and shunt malfunction.
- Watch for signs of infection, especially meningitis: fever, stiff neck, irritability, or tense fontanels. Also watch for redness, swelling, or

other signs of local infection over the shunt tract. Check dressing often for drainage.

- Listen for bowel sounds after ventriculoperitoneal shunt.
- Check the infant's growth and development periodically, and help the parents set goals consistent with ability and potential. Help parents focus on their child's strengths, not his weaknesses. Discuss special education programs, and emphasize the infant's need for sensory stimulation appropriate for his age. Teach parents to watch for signs of shunt malfunction, infection, and paralytic ileus. Tell them that surgery for lengthening the shunt will be required periodically as the child grows older. Surgery may also be required to correct shunt malfunctioning or to treat infection. Emphasize that hydrocephalus is a lifelong problem and that the child will require regular, continuing evaluation.

ARNOLD-CHIARI SYNDROME

Arnold-Chiari syndrome frequently accompanies hydrocephalus, especially when a myelomeningocele is also present. In this condition, an elongation or tonguelike downward projection of the cerebellum and medulla extends through the foramen magnum into the cervical portion of the spinal canal, impairing cerebrospinal fluid drainage from the fourth ventricle.

In addition to signs and symptoms of hydrocephalus, infants with Arnold-Chiari syndrome have nuchal rigidity, noisy respirations, irritability, vomiting, weak sucking reflex, and a preference for hyperextension of the neck.

Treatment requires surgery to insert a shunt like that used in hydrocephalus. Surgical decompression of the cerebellar tonsils at the foramen magnum is sometimes indicated.

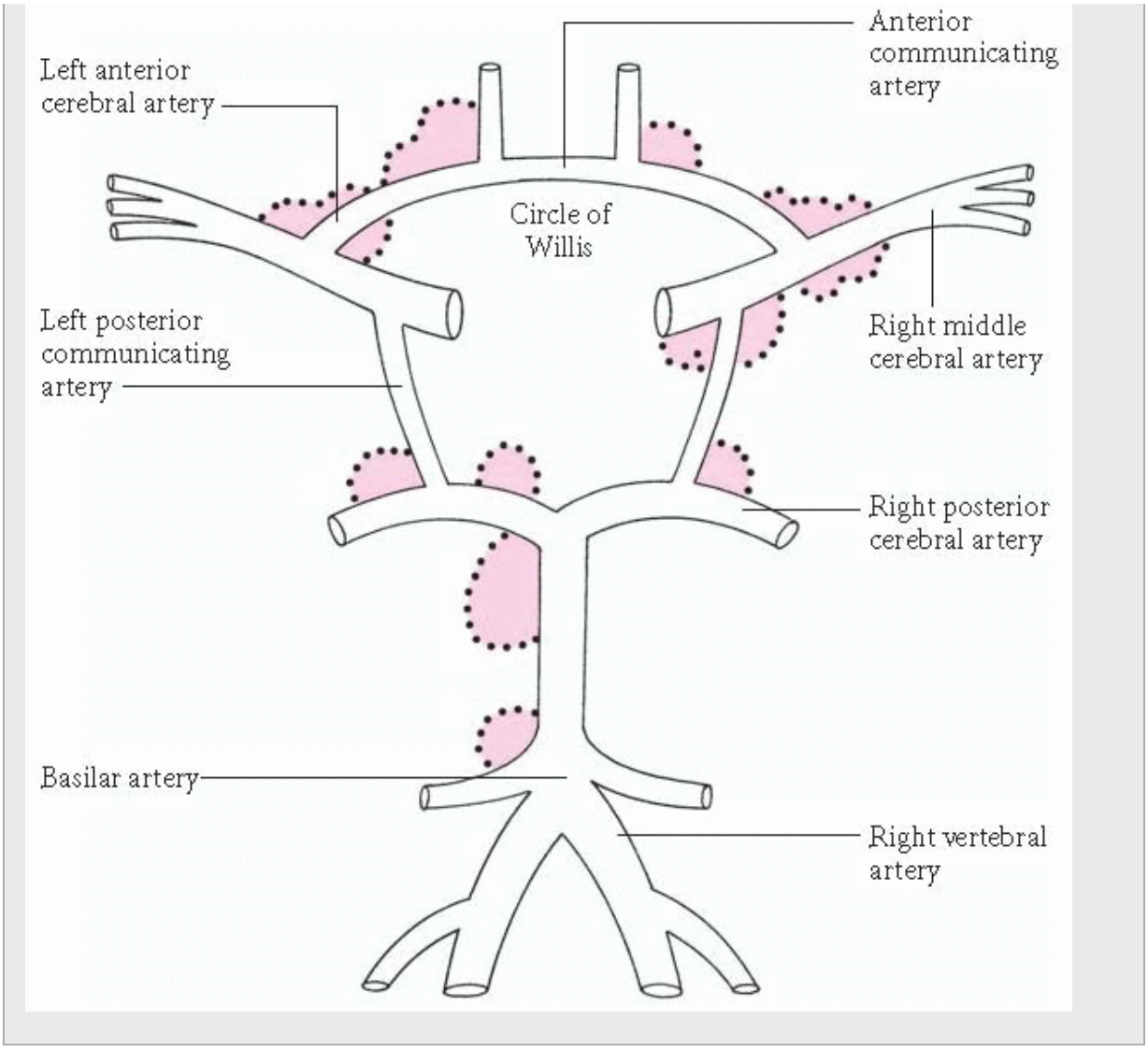
Cerebral aneurysm

Cerebral aneurysm is a localized dilation of a cerebral artery that typically results from a congenital weakness in the arterial wall. Its most common form is the berry aneurysm, a saclike outpouching in a cerebral artery. Cerebral aneurysms may arise at an

arterial junction in the Circle of Willis, the circular anastomosis forming the major cerebral arteries at the base of the brain. Cerebral aneurysms can rupture and cause subarachnoid hemorrhage. (See *Most common sites of cerebral aneurysm*.)

MOST COMMON SITES OF CEREBRAL ANEURYSM

Cerebral aneurysms usually arise at arterial bifurcations in the Circle of Willis and its branches. The illustration below shows the most common aneurysm sites around this circle.



The prognosis is guarded. Probably half the patients with subarachnoid hemorrhages die immediately; of those who survive untreated, 40% die from the effects of hemorrhage; another 20% die later from recurring hemorrhage. New treatments are improving the prognosis, however.

Causes and incidence

Cerebral aneurysm may result from a congenital defect, a degenerative process, or a combination of both. For example, hypertension and atherosclerosis may disrupt blood flow and exert pressure against a congenitally weak arterial wall, stretching it like an overblown balloon and making it likely to rupture. After such rupture, blood spills into the space normally occupied by cerebrospinal fluid (CSF), resulting in

subarachnoid hemorrhage. Blood may also spill into the brain tissue and form a clot, which can result in potentially fatal increased intracranial pressure (ICP) and brain-tissue damage.

Incidence is slightly higher in women than in men, especially those in their late 40s or early to mid-50s, but cerebral aneurysm may occur at any age, in both women and men.

Complications

(Most common after a rupture)

- Subarachnoid hemorrhage
- Brain tissue infarction
- Rebleeding
- Meningeal irritation
- Hydrocephalus

Signs and symptoms

Occasionally, rupture of a cerebral aneurysm causes premonitory symptoms that last several days, such as headache, nuchal rigidity, stiff back and legs, and intermittent nausea. Usually, however, onset is abrupt and without warning, causing a sudden severe headache, nausea, vomiting and, depending on the severity and location of bleeding, altered consciousness (including deep coma).

Bleeding causes meningeal irritation, resulting in nuchal rigidity, back and leg pain, fever, restlessness, irritability, occasional seizures, and blurred vision. Bleeding into the brain tissues causes hemiparesis, hemisensory defects, dysphagia, and visual defects. If the aneurysm is near the internal carotid artery, it compresses the oculomotor nerve and causes diplopia, ptosis, dilated pupil, and inability to rotate the eye.

The severity of symptoms varies considerably from patient to patient, depending on the site and amount of bleeding. To better describe their conditions, patients with ruptured cerebral aneurysms are grouped as follows:

- *Grade I (minimal bleed):* Patient is alert with no neurologic deficit; he may have a slight headache and nuchal rigidity.
- *Grade II (mild bleed):* Patient is alert, with a mild to severe headache, nuchal rigidity and, possibly, third-nerve palsy.
- *Grade III (moderate bleed):* Patient is confused or drowsy, with nuchal rigidity and, possibly, a mild focal deficit.
- *Grade IV (severe bleed):* Patient is stuporous, with nuchal rigidity and, possibly, mild to severe hemiparesis.
- *Grade V (moribund; commonly fatal):* If nonfatal, patient is in deep coma or decerebrate.

Generally, cerebral aneurysm poses three major threats:

- *Death from increased ICP:* Increased ICP may push the brain downward, impair brain stem function, and cut off blood supply to the part of the brain that supports vital functions.
- *Rebleed:* Generally, after the initial bleeding episode, a clot forms and seals the rupture, which reinforces the wall of the aneurysm for 7 to 10 days. However, after the 7th day, fibrinolysis begins to dissolve the clot and increases the risk of rebleeding. Signs and symptoms are similar to those accompanying the initial hemorrhage. Rebleeds during the first 24 hours after initial hemorrhage aren't uncommon, and they contribute to cerebral aneurysm's high mortality.
- *Vasospasm:* Why this occurs isn't clearly understood. Usually, vasospasm occurs in blood vessels adjacent to the cerebral aneurysm, but it may extend to major vessels of the brain, causing ischemia and altered brain function.

Other complications of cerebral aneurysm include pulmonary embolism (a possible adverse effect of deep vein thrombosis or aneurysm treatment) and acute hydrocephalus, occurring as CSF accumulates in the cranial cavity because of blockage by blood or adhesions.

Diagnosis

Diagnosis of cerebral aneurysm is based on the patient history and a neurologic examination; computed tomography scan, which reveals

subarachnoid or ventricular blood; or magnetic resonance imaging, which can identify a cerebral aneurysm as a flow void.

Cerebral angiography remains the procedure of choice for diagnosing cerebral aneurysm. Lumbar puncture may be used to identify blood in CSF if other studies are negative and the patient has no signs of increased ICP. Lumbar puncture should be performed if no contraindication is present and you strongly suspect a bleed, as imaging studies may miss a bleed.

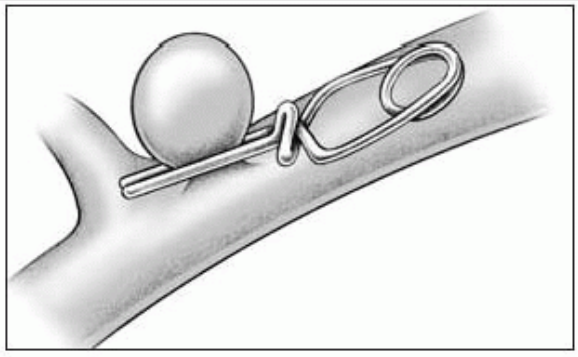
REPAIR OF CEREBRAL ANEURYSM

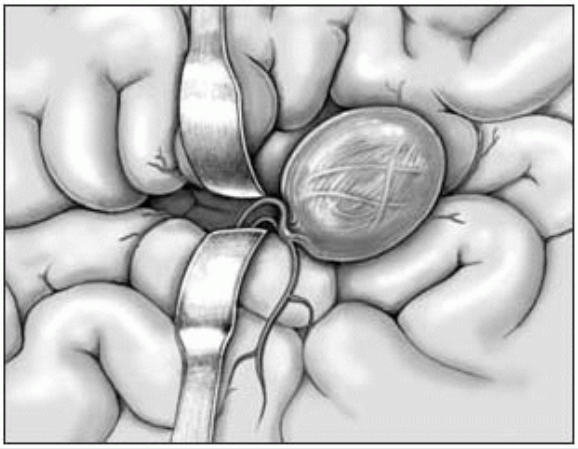
Clipping a cerebral aneurysm

The clip, which is made of materials that won't affect metal detectors and that will not rust, is placed at the base of the aneurysm to stop the blood supply. The clip remains in place permanently.

Coil embolization

In coil embolization, soft platinum coils are inserted into the aneurysm through the femoral artery. Usually 5 to 6 coils are needed to fill the aneurysm. The goal is to prevent blood flow into the aneurysm sac by filling the aneurysm with coils and thrombus.





Other baseline laboratory studies include complete blood count, urinalysis, arterial blood gas (ABG) analysis, coagulation studies, serum osmolality, and electrolyte and glucose levels.

Treatment

Coil embolization of the aneurysm aims to reduce the risk of bleeding, rebleeding after subarachnoid hemorrhage, and cerebral infarction or by clipping the aneurysm. (See *Repair of cerebral aneurysm*.) Both methods have advantages and disadvantages. Reinforcing the aneurysm with wrapping when the vessel is easily accessible is an alternative in specific situations. Surgical repair with clipping is usually performed at 7 to 10 days, whereas coil embolization has been found to be safe in the first 2 days after hemorrhage. Vasospasm, which previously had been the source of morbidity and mortality associated with successful surgical intervention, has shown therapeutic promise with transluminal balloon angioplasty and intra-arterial papaverine (Pava-Tine SR). In addition intra-arterial nimodipine, nicardipine (Cardene), verapamil (Isoptin) and milrinone (Primacor) as well as newer drugs fasudil hydrochloride and colforsin daropate are under investigation.

When surgical correction is delayed or hazardous due to aneurismal location or fitness of the patient, treatment includes:

- careful control of blood pressure (Calcium channel blockers are preferred.)
- bed rest and avoidance of head-down position

- avoidance of stimulants, including caffeine and catecholamines
- avoidance of platelet inhibitors such as aspirin
- corticosteroids (sometimes used to decrease edema but studies are not conclusive)
- anticonvulsants
- sedatives
- a fibrinolytic inhibitor, to minimize the risk of rebleed by delaying blood clot lysis.

After surgical repair, the patient's condition depends on the extent of damage from the initial bleed and the success of treatment for any resulting complications. Surgery can't improve the patient's neurologic condition unless it removes a hematoma or reduces the compression effect.

Special considerations

- An accurate neurologic assessment, good patient care, patient and family teaching, and psychological support can speed recovery and reduce complications.
- During initial treatment after hemorrhage, establish and maintain a patent airway if the patient needs supplementary oxygen. Position the patient to promote pulmonary drainage and prevent upper airway obstruction. If he's intubated, administering 100% oxygen before suctioning to remove secretions will prevent hypoxia and vasodilation from carbon dioxide accumulation. Suction no longer than 20 seconds to avoid increased ICP. Give frequent nose and mouth care.
- Impose aneurysm precautions to minimize the risk of rebleed and to avoid increased ICP. Such precautions include bed rest in a quiet, darkened room (keep the head of the bed flat or under 30 degrees, as ordered); limited visitors; avoiding strenuous physical activity and straining with bowel movements; and restricted fluid intake. Be sure to explain why these restrictive measures are necessary.

Preventive measures and good patient care can minimize other complications:

- Turn the patient often. Encourage occasional deep breathing and leg movement. Warn the patient to avoid all unnecessary physical activity. Assist with active range-of-motion exercises; if the patient is paralyzed, perform regular passive range-of-motion exercises.
- Monitor ABG levels, LOC, and vital signs often, and accurately measure intake and output. Avoid taking temperature rectally because vagus nerve stimulation may cause cardiac arrest.
- Watch for these danger signals, which may indicate an enlarging aneurysm, rebleeding, intracranial clot, vasospasm, or other complication: decreased LOC, unilateral enlarged pupil, onset or worsening of hemiparesis or motor deficit, increased blood pressure, slowed pulse, worsening of headache or sudden onset of a headache, renewed or worsened nuchal rigidity, and renewed or persistent vomiting. Intermittent signs such as restlessness, extremity weakness, and speech alterations can also indicate increasing ICP.
- Give fluids as ordered, and monitor I.V. infusions to avoid increased ICP.
- If the patient has facial weakness, assess the gag reflex and assist him during meals, placing food in the unaffected side of his mouth. If he can't swallow, insert a nasogastric tube as ordered, and give all tube feedings slowly. Prevent skin breakdown by taping the tube so it doesn't press against the nostril. If the patient can eat, provide a high-fiber diet to prevent straining at stool, which can increase ICP. Obtain an order for a stool softener, such as dioctyl sodium sulfosuccinate, or a mild laxative, and administer as ordered. Don't force fluids. Implement a bowel program based on previous habits. If the patient is receiving steroids, check the stool for blood.
- With third or facial nerve palsy, administer artificial tears or ointment to the affected eye, and tape the eye shut at night to prevent corneal damage.
- To minimize stress, encourage relaxation techniques. If possible, avoid using restraints because these can cause agitation and raise ICP.

- Administer antihypertensives as indicated. Carefully monitor blood pressure and immediately report *any* significant change, but especially a rise in systolic pressure.
- Prevent deep vein thrombosis by applying antiembolism stockings or sequential compression sleeves.
- If the patient can't speak, establish a simple means of communication, or use cards or a notepad. Try to limit conversation to topics that won't frustrate the patient. Encourage his family to speak to him in a normal tone, even if he doesn't seem to respond.
- Provide emotional support, and include the patient's family in his care as much as possible. Encourage family members to adopt a realistic attitude, but don't discourage hope.

- Before discharge, make a referral to a visiting nurse or a rehabilitation center when necessary, and teach the patient and his family how to recognize signs of rebleeding.

Arteriovenous malformations

Cerebral arteriovenous malformation (AVM) is a disorder of the blood vessels consisting of an abnormal connection between the arteries and the veins in the brain. It's a congenital disorder commonly resulting in tangled masses of thin-walled, dilated blood vessels between arteries and veins that aren't connected by capillaries. AVM primarily occurs in the posterior portion of the cerebral hemispheres. Adequate perfusion of brain tissue is prevented due to abnormal channels between the arterial and venous systems that allow mixing of oxygenated and unoxygenated blood. AVMs range in size from a few millimeters to large malformations that extend from the cerebral cortex to the ventricles. Patients typically present with multiple AVMs.

Complications of AVM include development of aneurysm and subsequent rupture, hemorrhage (intracerebral, subarachnoid, or subdural, depending on the location of the AVM), and hydrocephalus.

Causes and incidence

Although some AVMs occur as a result of penetrating injuries such as trauma, most are present at birth. However, symptoms typically don't occur until between the ages 10 and 20. Very large AVMs may short-circuit blood flow enough to cause cardiac decompensation, in which the heart can't pump enough blood to compensate for arteriovenous shunting in the brain. This typically occurs in infants and young children.

The vessels of an AVM are very thin and one or more arteries feed into it, causing it to appear dilated and tortuous. Typically, high-pressured arterial flow moves into the venous system through the connecting channels to increase venous pressure, engorging and dilating the venous structures. If the AVM is large enough, the shunting can deprive the surrounding tissue of adequate blood flow. Thin-walled vessels may ooze small amounts of blood—they may even rupture—causing hemorrhage into the brain or subarachnoid space.

Cerebral arteriovenous malformations occur in approximately 3 out of 10,000 people. Although the lesion is present at birth, symptoms may occur at any time. Two-thirds of cases occur before age 40. Evidence suggests that AVMs run in families. Males and females are affected equally.

Complications

- Aneurysm and subsequent rupture
- Intracerebral, subarachnoid, or subdural hemorrhage
- Hydrocephalus

Signs and symptoms

An AVM may be asymptomatic until complications occur; these may include rupture and a resulting sudden bleed in the brain, known as a hemorrhagic stroke. Arteriovenous malformations vary in size and location within the brain. Systolic bruit may be auscultated over the carotid artery, mastoid process, or orbit on examination.

Symptoms that occur prior to an AVM rupture are related to smaller and slower bleeding from the abnormal vessels, which are usually fragile because their structure is abnormal.

In more than half of patients with AVM, hemorrhage from the malformation is the first symptom. Depending on the location and the severity of the bleed, the hemorrhage can be profoundly disabling or fatal. The risk of bleeding from an AVM is approximately 2% to 4% per year.

The first symptoms often include headache, seizure, or other sudden neurological problems, such as vision problems, weakness, inability to move a limb or a side of the body, lack of sensation in part of the body, or abnormal sensations, such as ringing and numbness. Symptoms are the same as for stroke. The individual with an AVM may complain of chronic mild headache, a sudden and severe headache, or a localized or general headache. The headache may resemble migraine and vomiting may occur. Seizures may result from focal neurologic

deficits (depending on the location of the AVM) resulting from compression and diminished perfusion. Symptoms of intracranial (intracerebral, subarachnoid, or subdural) hemorrhage result. Muscle weakness and decreased sensation can occur in any part of the body. Mental status change can occur where the individual appears sleepy, stuporous, lethargic, confused, disoriented, or irritable. Additional symptoms may include stiff neck, speech or sense of smell impairment, dysfunctional movement, fainting, facial paralysis, eyelid drooping, tinnitus, dizziness, and decreased level of consciousness (LOC).

If an AVM bleeds once, the risk is greater that it will bleed again in the future. Intracerebral or subarachnoid hemorrhages are the most common first symptoms of cerebral arteriovenous malformation. In some cases, symptoms may also occur due to lack of blood flow to an area of the brain (ischemia), compression or distortion of brain tissue by large AVMs, or abnormal brain development in the area of the malformation. Progressive loss of nerve cells in the brain may occur, caused by mechanical (pressure) and ischemic (lack of blood supply) factors.

Diagnosis

Tests used to diagnose AVM include head computed tomography scan, cranial magnetic resonance imaging, and magnetic resonance angiography. An EEG may be performed if symptoms include seizures, but this test isn't diagnostic of the specific area of the lesion.

Cerebral arteriogram confirms the presence of AVMs and evaluates blood flow. Doppler ultrasonography of cerebrovascular system indicates abnormal, turbulent blood flow.

Treatment

General support measures include aneurysm precautions to prevent possible rupture. This involves placing the patient on bed rest or with limited activity and maintaining a quiet atmosphere. Analgesics may be given for headache, and sedatives may be given to help calm the patient and prevent rupture. Stool softeners may be given to prevent straining at stool, which increases intracranial pressure.

A bleeding AVM is a medical emergency requiring immediate hospitalization. The goal of treatment is to prevent further complications by limiting bleeding, controlling seizures and, if possible, removing the AVM. Surgery for correction may include block dissection, laser, or ligation to repair the communicating channels and remove the feeding vessels. Embolization or radiation therapy may be done, if surgery isn't possible, to close the communicating channels and feeder vessels, thereby reducing blood flow to the AVM. Open brain surgery, endovascular treatment, and radiosurgery may be used separately or in any combination, depending upon the physician and the patient's individual situation. Surgery is dependent upon the accessibility and size of the lesion and the patient's status. Open brain surgery involves the actual removal of the malformation in the brain through an opening made in the skull. This surgery is particularly risky because the surgery itself may cause the AVM to bleed uncontrollably.

Embolization (injecting a gluelike substance into the abnormal vessels to stop aberrant blood flow into the AVM) may be an alternative if surgery isn't feasible due to the size or location of the lesion. Stereotactic radiosurgery may also be an alternative for patients with inoperable arteriovenous malformations. It's particularly useful for small, deep lesions, which are difficult to remove by surgery.

Anticonvulsant medications such as phenytoin are usually prescribed if seizures occur.

Special considerations

- Monitor vital signs and adjust medications to control hypertension.
- Monitor neurologic status.
- Monitor for seizure activity and institute seizure precautions.
- Maintain a quiet atmosphere and provide relaxation techniques.
- Discuss the importance of reporting any signs of intracranial bleeding immediately (sudden severe headache, vision changes, decreased movement in extremities, and change in LOC).

- Refer to social service for support services if neurological deficits have occurred from a ruptured AVM.

PAROXYSMAL DISORDERS

Headache

The most common patient complaint, headache usually occurs as a symptom of an underlying disorder. Ninety percent of all headaches are vascular, muscle contraction, or a combination; 10% are due to underlying intracranial, systemic, or psychological disorders. Migraine headaches, probably the most intensively studied, are throbbing, vascular headaches that usually begin to appear in childhood or adolescence and recur throughout adulthood.

Causes and incidence

Most chronic headaches result from tension (muscle contraction), which may be caused by emotional stress, fatigue, menstruation, or environmental stimuli (noise, crowds, or bright lights). Other possible causes include glaucoma; inflammation of the eyes or mucosa of the nasal or paranasal sinuses; diseases of the scalp, teeth, extracranial arteries, or external or middle ear; muscle spasms of the face, neck, or shoulders; and cervical arthritis. In addition, headaches may be caused by vasodilators (nitrates, alcohol, and histamine), systemic disease, hypoxia, hypertension, head trauma and tumor, intracranial bleeding, abscess, or aneurysm.

The cause of migraine headache is unknown, but it's associated with constriction and dilation of intracranial and extracranial arteries. Certain biochemical abnormalities are thought to occur during a migraine attack. These include local leakage of a vasodilator polypeptide called neurokinin through the dilated arteries and a decrease in the plasma level of serotonin.

Headache pain may emanate from the pain-sensitive structures of the skin, scalp, muscles, arteries, and veins; cranial nerves V, VII, IX, and X; or cervical nerves 1, 2, and 3. Intracranial mechanisms of headaches include traction or displacement of arteries, venous sinuses, or venous tributaries and inflammation or direct pressure on the cranial nerves with afferent pain fibers.

Affecting up to 10% of Americans, headaches are more common in females and have a strong familial incidence. Drops in estrogen level may precipitate migraine headaches.

Complications

- Worsening of preexisting hypertension
- Photophobia
- Emotional lability
- Motor weakness

Signs and symptoms

Initially, migraine headaches usually produce unilateral, pulsating pain, which later becomes more generalized. They're commonly preceded by a scintillating scotoma, hemianopsia, unilateral paresthesia, or speech disorders. The patient may experience irritability, anorexia, nausea, vomiting, and photophobia. (See *Clinical features of migraine headaches*.)

Both muscle contraction and tractioninflammatory vascular headaches produce a dull, persistent ache, tender spots on the head and neck, and a feeling of tightness around the head, with a characteristic “hatband” distribution. The pain is usually severe and unrelenting. If caused by intracranial bleeding, these headaches may result in neurologic deficits,

such as paresthesia and muscle weakness; narcotics may fail to relieve pain in these cases. If caused by a tumor, pain is most severe when the patient awakens.

Diagnosis

Diagnosis requires a history of recurrent headaches and physical examination of the head and neck. Such examination includes percussion, auscultation for bruits, inspection for signs of infection, and palpation for defects, crepitus, or tender spots (especially after trauma). Firm diagnosis also requires a complete neurologic examination, assessment for other systemic diseases—such as hypertension—and a psychosocial

evaluation, when such factors are suspected.

CLINICAL FEATURES OF MIGRAINE HEADACHES

Type	Signs and symptoms
Common migraine (most prevalent)	
Usually occurs on weekends and holidays	▪ Prodromal symptoms (fatigue, nausea, vomiting, and fluid imbalance) precede headache by about 1 day. ▪ Sensitivity to light and noise (most prominent feature) ▪ Headache pain (unilateral or bilateral, aching or throbbing)
Classic migraine	
Usually occurs in compulsive personalities and within families	▪ Prodromal symptoms include visual disturbances, such as zigzag lines and bright lights (most common), sensory disturbances (tingling of face, lips, and hands), or motor disturbances (staggering gait). ▪ Recurrent and periodic headaches
Hemiplegic and ophthalmoplegic migraine (rare)	
Usually occurs in young adults	▪ Severe, unilateral pain ▪ Extraocular muscle palsies (involving third cranial nerve) and ptosis

	▪ With repeated headaches, possible permanent third cranial nerve injury ▪ In hemiplegic migraine, neurologic deficits (hemiparesis, hemiplegia) may persist after the headache subsides.
Basilar artery migraine	
Occurs in young women before their menstrual periods	▪ Prodromal symptoms usually include partial vision loss followed by vertigo, ataxia, dysarthria, tinnitus and, sometimes, tingling of the fingers and toes, lasting from several minutes to almost an hour. ▪ Headache pain, severe occipital throbbing, vomiting
Cluster headaches	
Occur in men more commonly than women and occur at all ages, but more commonly in adolescents and middle-age people.	▪ Episodic type (more common) and involves one to three short-lived attacks of periorbital pain per day over a 4- to 8-week period followed by a pain-free interval averaging 1 year. Chronic type occurs after an episodic pattern is established. ▪ Unilateral pain occurs without warning, reaching a crescendo within 5 minutes, and described as excruciating and deep, with attacks lasting from 30 minutes to 2 hours. ▪ Associated symptoms may include tearing, reddening of the eye, nasal stuffiness, lid ptosis, and nausea.

Diagnostic tests include cervical spine and sinus X-rays, EEG, computed tomography scan—performed before lumbar puncture to rule out increased intracranial pressure (ICP)—or magnetic resonance imaging. A lumbar puncture isn't done if there's evidence of increased ICP or if a brain tumor is suspected because rapidly

reducing pressure by removing spinal fluid can cause brain herniation.

Treatment

Depending on the type of headache, analgesics —ranging from aspirin to codeine or meperidine—may provide symptomatic relief. Other measures include identification and elimination of causative factors and, possibly,

psychotherapy for headaches caused by emotional stress. Chronic tension headaches may also respond to muscle relaxants.

For migraine headaches, ergotamine alone or with caffeine may be an effective treatment. The Food and Drug Administration allows labeling of various analgesic preparations that include caffeine to state that they're for the treatment of migraine headaches. Remember that these medications can't be taken by pregnant women because they stimulate uterine contractions. These drugs and others, such as metoclopramide or non-steroidal anti-inflammatories, work best when taken early in the course of an attack. If nausea and vomiting make oral administration impossible, drugs may be given as rectal suppositories.

Drugs in the class of sumatriptan are considered by many clinicians to be the drug of choice for acute migraine attacks or cluster headaches. Drugs that can help prevent migraine headaches include antidepressants (such as nortriptyline or fluoxetine), beta blockers (propranolol), and calcium-channel blockers (verapamil). Corticosteroids provide short-term relief for some patients with cluster headaches.

Special considerations

- Headaches seldom require hospitalization unless caused by a serious disorder. If that's the case, direct your care to the underlying problem.
- Obtain a complete patient history: duration and location of the headache; time of day it usually begins; nature of the pain; concurrence with other symptoms such as blurred vision; precipitating factors, such as tension, menstruation, loud noises, menopause, or alcohol; medications taken such as oral contraceptives; or prolonged fasting. Exacerbating factors can also be assessed through ongoing observation of the patient's personality, habits, activities of daily living, family relationships, coping mechanisms, and relaxation activities.
- Using the history as a guide, help the patient avoid exacerbating factors. Advise him to lie down in a dark, quiet room during an attack and to place ice packs on his forehead or a cold cloth over his eyes.
- Instruct the patient to take the prescribed medication at the onset of migraine symptoms, to prevent dehydration by drinking plenty of

fluids after nausea and vomiting subside, and to use other headache relief measures.

- The patient with a migraine headache usually needs to be hospitalized only if nausea and vomiting are severe enough to induce dehydration and possible shock.
- Avoid repeated use of narcotics if possible.

PREVENTION

Advise patient to:

- *get adequate sleep*
- *eat a well-balanced diet and drink plenty of water*
- *do upper-body stretching exercises.*
- *quit smoking*
- *learn relaxation techniques such as yoga, medication, and deep breathing*
- *avoid known causative triggers.*

Seizure disorder

Seizure disorder, also called epilepsy, is a condition of the brain marked by a susceptibility to recurrent seizures—paroxysmal events associated with abnormal electrical discharges of neurons in the brain.

Causes and incidence

In about half the cases of seizure disorder, the cause is unknown. However, some possible causes of seizure disorder include:

- birth trauma (inadequate oxygen supply to the brain, blood incompatibility, or hemorrhage)
- perinatal infection
- anoxia (after respiratory or cardiac arrest)
- infectious diseases (meningitis, encephalitis, or brain abscess)

- ingestion of toxins (mercury, lead, or carbon monoxide)
- tumors of the brain
- inherited disorders or degenerative disease, such as phenylketonuria or tuberous sclerosis
- head injury or trauma
- metabolic disorders, such as hypoglycemia or hypoparathyroidism
- stroke (hemorrhage, thrombosis, or embolism).

Alcohol withdrawal can cause nonepileptic seizures.

Seizure disorder affects 1% to 2% of the population. However, 80% of patients have good seizure control if they strictly adhere to the prescribed treatment regimen.

Complications

- Anoxia (during a seizure)
- Fall injuries

Signs and symptoms

The hallmarks of seizure disorder are recurring seizures, which can be classified as partial or generalized (some patients may be affected by more than one type).

Partial seizures arise from a localized area of the brain, causing specific symptoms. In some patients, partial seizure activity may spread to the entire brain, causing a generalized seizure. Partial seizures include simple partial (jacksonian) and complex partial seizures (psychomotor or temporal lobe).

A simple partial motor-type seizure begins as a localized motor seizure characterized by a spread of abnormal activity to adjacent areas of the brain. It typically produces stiffening or jerking in one extremity, accompanied by a tingling sensation in the same area. For example, it may start in the thumb and spread to the entire hand and arm. The

patient seldom loses consciousness, although the seizure may progress to a generalized seizure.

A simple partial sensory-type seizure involves perceptual distortion, which can include hallucinations.

The symptoms of a complex partial seizure vary but usually include purposeless behavior. The patient experiences an aura immediately before the seizure. An aura represents the beginning of abnormal electrical discharges within a focal area of the brain and may include a pungent smell, GI distress (nausea or indigestion), a rising or sinking feeling in the stomach, a dreamy feeling, an unusual taste, or a visual disturbance. Overt signs of a complex partial seizure include a glassy stare, picking at one's clothes, aimless wandering, lipsmacking or chewing motions, and unintelligible speech; these signs may last for just a few seconds or as long as 20 minutes. Mental confusion may last several minutes after the seizure; as a result, an observer may mistakenly suspect intoxication with alcohol or drugs or psychosis.

Generalized seizures, as the term suggests, cause a generalized electrical abnormality within the brain and include several distinct types:

- Absence *(petit mal)* seizures occur most commonly in children, although they may affect adults as well. They usually begin with a brief change in level of consciousness, indicated by blinking or rolling of the eyes, a blank stare, and slight mouth movements. There's little or no tonic-clonic movement. The patient retains his posture and continues preseizure activity without difficulty. Typically, each seizure lasts from 1 to 10 seconds. If not properly treated, seizures can recur as often as 100 times per day. An absence seizure may progress to generalized tonic-clonic seizures.
- A myoclonic *(bilateral massive epileptic myoclonus)* seizure is characterized by brief, involuntary muscular jerks of the body or extremities, which may occur in a rhythmic fashion and may precede generalized tonic-clonic seizures by months or years.
- A generalized tonic-clonic *(grand mal)* seizure typically begins with a loud cry, precipitated by air rushing from the lungs through the vocal cords. The patient then falls to the ground, losing consciousness. The body stiffens (tonic phase) and then alternates between episodes of

muscular spasm and relaxation (clonic phase). Tongue-biting, incontinence, labored breathing, apnea, and subsequent cyanosis may also occur. The seizure stops in 2 to 5 minutes, when abnormal electrical conduction

of the neurons is completed. The patient then regains consciousness but is somewhat confused and may have difficulty talking. If he can talk, he may complain of drowsiness, fatigue, headache, muscle soreness, and arm or leg weakness. He may fall into deep sleep after the seizure. These seizures may start as facial seizures and spread to become generalized.

An akinetic seizure is characterized by a general loss of postural tone (the patient falls in a flaccid state) and a temporary loss of consciousness. It occurs in young children and is sometimes called a “drop attack” because it causes the child to fall.

Status epilepticus is a continuous seizure state that can occur in all seizure types. The most life-threatening example is generalized tonic-clonic status epilepticus, a continuous generalized tonic-clonic seizure without intervening return of consciousness. Status epilepticus is accompanied by respiratory distress. It can result from abrupt withdrawal of anticonvulsant medications, hypoxic encephalopathy, acute head trauma, metabolic encephalopathy, or septicemia secondary to encephalitis or meningitis.

Diagnosis

Clinically, the diagnosis of seizure disorder is based on the occurrence of one or more seizures and proof or the assumption that the condition that led to them is still present.

Diagnostic information is obtained from the patient's history and description of seizure activity and from family history, physical and neurologic examinations, and computed tomography scan or magnetic resonance imaging. These scans offer density readings of the brain and may indicate abnormalities in internal structures. Paroxysmal abnormalities on the EEG confirm the diagnosis by providing evidence of the continuing tendency to have seizures. A negative EEG doesn't rule out seizure disorder because the paroxysmal abnormalities occur

intermittently. Other tests may include serum glucose and calcium studies, skull X-rays, lumbar puncture, brain scan, and cerebral angiography.

Treatment

Generally, treatment of seizure disorder consists of anticonvulsant therapy to reduce the number of future seizures. The most commonly prescribed drugs include phenytoin, carbamazepine, phenobarbital, gabapentin, or primidone administered individually for generalized tonic-clonic seizures and complex partial seizures. Valproic acid, clonazepam, and ethosuximide are commonly prescribed for absence seizures. Gabapentin and felbamate are also anticonvulsant drugs.

A patient taking anticonvulsant medications requires monitoring for toxic signs: nystagmus, ataxia, lethargy, dizziness, drowsiness, slurred speech, irritability, nausea, and vomiting.

If drug therapy fails, treatment may include surgical removal of a demonstrated focal lesion to attempt to stop seizures. Emergency treatment of status epilepticus usually consists of diazepam (Valium) or lorazepam (Ativan), phenytoin or phenobarbital (Solfoton); dextrose 50% I.V. (when seizures are secondary to hypoglycemia); and thiamine I.V. (in chronic alcoholism or withdrawal).

Special considerations

A key to support is a true understanding of the nature of seizure disorder and of the misconceptions that surround it.

- Encourage the patient and his family to express their feelings about the patient's condition. Answer their questions, and help them cope by dispelling some of the myths about seizure disorder, for example, the myth that seizure disorder is contagious. Assure them that seizure disorder is controllable for most patients who follow a prescribed regimen of medication and that most patients maintain a normal lifestyle.

Because drug therapy is the treatment of choice for most people with seizure disorder, information about medications is invaluable.

- Stress the need for compliance with the prescribed drug schedule. Reinforce dosage instructions and stress the importance of taking medication regularly, at scheduled times. Caution the patient to monitor the quantity of medication he has so he doesn't run out of it.

- Warn against possible adverse effects—drowsiness, lethargy, hyperactivity, confusion, and visual and sleep disturbances—all of which indicate the need for dosage adjustment. Phenytoin therapy may lead to hyperplasia of the gums, which may be relieved by conscientious oral hygiene. Instruct the patient to report adverse effects immediately.
- When administering phenytoin I.V., use a large vein and monitor vital signs frequently or administer fosphenytoin (Cerebyx), which has fewer vascular side effects. Avoid I.M. administration and mixing with dextrose solutions.
- Emphasize the importance of having anticonvulsant blood levels checked at regular intervals, even if the seizures are under control.
- Warn the patient against drinking alcoholic beverages.
- Know which social agencies in your community can help epileptic patients. Refer the patient to the Epilepsy Foundation of America for general information and to the state motor vehicle department for information about a driver's license.

The primary goals of the health care professional and family members caring for a patient having a seizure are protection from injury, protection from aspiration, and observation of the seizure activity. Generalized tonic-clonic seizures may necessitate first aid. Show the patient's family members how to administer first aid correctly:

- Avoid restraining the patient during a seizure. Help the patient to a lying position, loosen any tight clothing, and place something flat and soft, such as a pillow, jacket, or hand, under his head. Clear the area of hard objects. *Don't* force anything into the patient's mouth if his teeth are clenched—a tongue blade or spoon could lacerate a mouth and lips or displace teeth, precipitating respiratory distress. However, if the patient's mouth is open, protect his tongue by placing a soft object (such as a folded cloth) between his teeth. Turn his head to

provide an open airway. After the seizure subsides, reassure the patient that he's all right, orient him to time and place, and inform him that he's had a seizure.

- *Don't* restrain the patient during a complex partial seizure. Clear the area of any hard objects. Protect him from injury by gently calling his name and directing him away from the source of danger. After the seizure passes, reassure him and tell him that he has just had a seizure.

BRAIN AND SPINAL CORD DISORDERS

Stroke

A stroke, also called *cerebrovascular accident* or *brain attack,* is a sudden impairment of cerebral circulation in one or more of the blood vessels supplying the brain. A stroke interrupts or diminishes oxygen supply and commonly causes serious damage or necrosis in brain tissues. The sooner circulation returns to normal after a stroke, the better chances are for complete recovery. However, about half of those who survive a stroke remain permanently disabled and experience a recurrence within weeks, months, or years.

Causes and incidence

A stroke results from obstruction of a blood vessel, typically in extracerebral vessels, but occasionally in intracerebral vessels. Factors that increase the risk of stroke include history of transient ischemic attacks (TIAs), atherosclerosis, hypertension, kidney disease, arrhythmias (specifically atrial fibrillation), electrocardiogram changes, rheumatic heart disease, diabetes mellitus, postural hypotension, cardiac or myocardial enlargement, high serum triglyceride levels, lack of exercise, use of oral contraceptives, cigarette smoking, and family history of stroke. (See *Transient ischemic attack*, page 192.)

The major causes of stroke are thrombosis, embolism, and hemorrhage. Thrombosis is the most common cause in middle-age and elderly people, who have a higher incidence of atherosclerosis, diabetes, and hypertension. Thrombosis causes ischemia in brain tissue supplied by the affected vessel as well as congestion and edema; the

latter may produce more clinical effects than thrombosis itself, but these symptoms subside with the edema. Thrombosis may develop while the patient sleeps or shortly after he awakens; it can also occur during surgery or after a myocardial infarction. The risk increases with obesity, smoking, or the use of oral contraceptives. Cocaineinduced ischemic stroke is now being seen in younger patients.

TRANSIENT ISCHEMIC ATTACK

A transient ischemic attack (TIA) is a recurrent episode of neurologic deficit, lasting from seconds to hours, that clears within 12 to 24 hours. It's usually considered a warning sign of an impending thrombotic stroke. In fact, TIAs have been reported in 50% to 80% of patients who have had a cerebral infarction from such thrombosis. The age of onset varies. Incidence rises dramatically after age 50 and is highest among blacks and men.

Causes

In TIA, microemboli released from a thrombus probably temporarily interrupt blood flow, especially in the small distal branches of the arterial tree in the brain. Small spasms in those arterioles may impair blood flow and also precede TIA. Predisposing factors are the same as for thrombotic strokes. The most distinctive characteristics of TIAs are the transient duration of neurologic deficits and complete return of normal function. The symptoms of TIA easily correlate with the location of the affected artery. These symptoms include double vision, speech deficits (slurring or thickness), unilateral blindness, staggering or uncoordinated gait, unilateral weakness or numbness, falling because of weakness in the legs, and dizziness.

Treatment

During an active TIA, treatment aims to prevent a completed stroke and consists of aspirin or

anticoagulants to minimize the risk of thrombosis. After or between attacks, preventive treatment includes carotid endartectomy or cerebral microvascular bypass.

Embolism, the second most common cause of stroke, is an occlusion of a blood vessel caused by a fragmented clot, a tumor, fat, bacteria, or air. It can occur at any age, especially among patients with a history of rheumatic heart disease, endocarditis, posttraumatic valvular disease, myocardial fibrillation and other cardiac arrhythmias, or after open-heart surgery. It usually develops rapidly—in 10 to 20 seconds—and without warning. When an embolus reaches the cerebral vasculature, it cuts off circulation by lodging in a narrow portion of an artery, most commonly the middle cerebral artery, causing necrosis and edema. If the embolus is septic and infection extends beyond the vessel wall, encephalitis or an abscess may develop.

Hemorrhage, the third most common cause of stroke, may, like embolism, occur suddenly, at any age, and affects more women than men. Hemorrhage results from chronic hypertension or aneurysms, which cause sudden rupture of a cerebral artery, thereby diminishing blood supply to the area served by the artery. In addition, blood accumulates deep within the brain, further compressing neural tissue and causing even greater damage.

Strokes are classified according to their course of progression. The least severe is the TIA, or “little stroke,” which results from a temporary interruption of blood flow, most commonly in the carotid and vertebrobasilar arteries. A progressive stroke, or stroke-in-evolution (thrombusin-evolution), begins with slight neurologic deficit and worsens in a day or two. In a completed stroke, neurologic deficits are maximal at onset.

Stroke is the third most common cause of death in most developed countries today and the most common cause of neurologic disability. It occurs in 1 out of 15 deaths in the United States each year.

Complications

- Unstable blood pressure

- Fluid imbalances
- Infection
- Sensory impairment
- Altered level of consciousness
- Aspiration
- Pulmonary emboli

Signs and symptoms

Clinical features of stroke vary with the artery affected (and, consequently, the portion of the brain it supplies), the severity of damage, and the extent of collateral circulation that develops to help the brain compensate for decreased blood supply. If the stroke occurs in the left hemisphere, it produces symptoms on the right side; if in the right hemisphere, symptoms are on the left side. However, a stroke that causes cranial nerve damage produces signs of cranial nerve dysfunction on the same side as the hemorrhage.

Symptoms are usually classified according to the artery affected:

- *Middle cerebral artery:* aphasia, dysphasia, visual field cuts, and hemiparesis on affected side (more severe in the face and arm than in the leg)
- *Carotid artery:* weakness, paralysis, numbness, sensory changes, and visual disturbances on affected side; altered level of consciousness (LOC), bruits, headaches, aphasia, and ptosis
- *Vertebrobasilar artery:* weakness on affected side, numbness around lips and mouth, visual field cuts, diplopia, poor coordination, dysphagia, slurred speech, dizziness, amnesia, and ataxia
- *Anterior cerebral artery:* confusion, weakness, and numbness (especially in the leg) on affected side, incontinence, loss of coordination, impaired motor and sensory functions, and personality changes
- *Posterior cerebral arteries:* visual field cuts, sensory impairment, dyslexia, coma, and cortical blindness; typically, paralysis is absent.

Symptoms can also be classified as premonitory, generalized, and focal. Premonitory symptoms, such as drowsiness, dizziness, headache, and mental confusion, are rare. Generalized symptoms, such as headache, vomiting, mental impairment, seizures, coma, nuchal rigidity, fever, and disorientation, are typical. Focal symptoms, such as sensory and reflex changes, reflect the site of hemorrhage or infarct and may worsen.

Diagnosis

Diagnosis of stroke is based on observation of clinical features, a history of risk factors, and the results of diagnostic tests:

- Computed tomography scan shows evidence of hemorrhagic stroke immediately but may not show evidence of thrombotic infarction for 48 to 72 hours.
- Magnetic resonance imaging may help identify ischemic or infarcted areas and cerebral swelling.
- Electrocardiogram can help diagnose underlying heart disorders.
- Carotid duplex may detect carotid artery stenosis.
- Angiography outlines blood vessels and pinpoints occlusion or rupture site. It's mainly used if surgery is considered.
- EEG helps to localize damaged area.

Other baseline laboratory studies may be done to exclude immune conditions or abnormal clotting that can lead to clot formation.

Treatment

Surgery performed to improve cerebral circulation for patients with thrombotic or embolic stroke includes endarterectomy (removal of atherosclerotic plaques from the inner arterial wall) and microvascular bypass (surgical anastomosis of an extracranial vessel to an intracranial vessel).

Medications useful in stroke include:

- Tissue plasminogen activator has been used successfully in clot dissolution when administered within 3 hours of the onset of

symptoms. In other circumstances, heparin and warfarin (Coumadin) may be used, as well as aspirin and other antiplatelet drugs.

- Ticlopidine, an antiplatelet drug, may be more effective than aspirin in preventing stroke and reducing the risk of recurrent stroke after therapy has begun.
- Anticonvulsants may be used to treat or prevent seizures.

- Stool softeners may be used to prevent straining, which increases intracranial pressure (ICP).
- Corticosteroids may be indicated to minimize associated cerebral edema.
- Analgesics may be used to relieve the headache that typically follows hemorrhagic stroke.

Special considerations

During the acute phase, efforts focus on survival needs and prevention of further complications. Effective care emphasizes continuing neurologic assessment, support of respiration, continuous monitoring of vital signs, careful positioning to prevent aspiration and contractures, management of GI problems, and careful monitoring of fluid, electrolyte, and nutritional status. Patient care must also include measures to prevent complications such as infection.

- Maintain a patent airway and oxygenation. Loosen constricting clothes. Watch for ballooning of the cheek with respiration. The side that balloons is the side affected by the stroke. If the patient is unconscious, he could aspirate saliva, so keep him in a lateral position to allow secretions to drain naturally or suction secretions, as needed. Insert an artificial airway, and start mechanical ventilation or supplemental oxygen, if necessary.
- Check vital signs and neurologic status, record observations, and report any significant changes to the physician. Monitor blood pressure, LOC, pupillary changes, motor function (voluntary and involuntary movements), sensory function, speech, skin color, temperature, signs of increased ICP, and nuchal rigidity or flaccidity.

Remember, if stroke is impending, blood pressure rises suddenly, pulse is rapid and bounding, and the patient may complain of a headache. Also, watch for signs of pulmonary emboli, such as chest pains, shortness of breath, dusky color, tachycardia, fever, and changed sensorium. If the patient is unresponsive, monitor his blood gases often and alert the physician to increased partial pressure of carbon dioxide or decreased partial pressure of oxygen.

- Maintain fluid and electrolyte balance. If the patient can take liquids orally, offer them as often as fluid limitations permit. Administer I.V. fluids as ordered; never give too much too fast because this can increase ICP. Offer the urinal or bedpan every 2 hours. If the patient is incontinent, he may need an indwelling urinary catheter, but this should be avoided, if possible, because of the risk of infection.
- Ensure adequate nutrition. Check for gag reflex before offering small oral feedings of semisolid foods. Place the food tray within the patient's visual field because loss of peripheral vision is common. If oral feedings aren't possible, insert a nasogastric tube.
- Manage GI problems. Be alert for signs that the patient is straining at elimination because this increases ICP. Modify diet, administer stool softeners, as indicated, and give laxatives, if necessary. If the patient vomits (usually during the first few days), keep him positioned on his side to prevent aspiration.
- Provide careful mouth care. Clean and irrigate the patient's mouth to remove food particles. Care for his dentures as needed.
- Provide meticulous eye care. Remove secretions with a cotton ball and sterile normal saline solution. Instill eyedrops as ordered. Patch the patient's affected eye if he can't close the lid.
- Position the patient and align his extremities correctly. Use high-topped sneakers to prevent footdrop and contracture and convoluted foam, flotation, or pulsating mattresses, or sheepskin, to prevent pressure ulcers. To prevent pneumonia, turn the patient at least every 2 hours. Elevate the affected hand to control dependent edema, and place it in a functional position.
- Assist the patient with exercise. Perform range-of-motion exercises for both the affected and unaffected sides. Teach and encourage the

patient to use his unaffected side to exercise his affected side.

- Give medications as ordered, and watch for and report adverse effects.
- Establish and maintain communication with the patient. If he is aphasic, set up a simple method of communicating basic needs. Then, remember to phrase your questions so he'll be able to answer using this system. Repeat yourself quietly and calmly, and use gestures if necessary to

help him understand. Even an unresponsive patient may be able to hear, so don't say anything in his presence you wouldn't want him to hear and remember.

- Provide psychological support. Set realistic short-term goals. Involve the patient's family in his care when possible, and explain his deficits and strengths.
- If the patient has a visual field deficit, make sure caregivers and family members approach the patient from his visually intact side.

PREVENTION

PREVENTING STROKE

Lifestyle modifications can help prevent heart attack and stroke. The American Heart Association recommends the following:

Healthy diet

Give patients the following advice:

- Eat five or more servings of fruit and vegetables daily.
- Eat six or more servings a day of grain products including whole grains.
- Eat fish at least twice a week, especially mackerel, lake trout, herring, sardines, albacore tuna and salmon.
- Include fat-free and low-fat milk products, beans, lean meats, and skinless poultry.

- Choose fats and oils with 2 grams or less of saturated fat per serving (1 tablespoon).
- Limit your intake of foods high in calories or low in protein, such as carbonated beverages, high-sugar foods, and candy.
- Eat less than 6 grams of salt per day (1 teaspoon)
- Limit foods high in saturated fat, trans fat, or cholesterol, such as whole milk, fatty meats, tropical oils, and partially hydrogenated vegetable oils.

Exercise

Regular physical activity is defined by the American Heart Association as moderate to vigorous exercise 30 minutes a day on most or all days of the week. A lack of physical activity can lead to obesity and increase the risk of hypertension, heart attack, and stroke.

Smoking cessation

Even smoking filtered and light or ultralight cigarettes can lead to atherosclerosis. Quitting or not starting is the only thing that can prevent this major risk factor for heart attack and stroke.

Blood pressure awareness

Individuals should know what their blood pressure is by having it checked by a practitioner. If it's high, it may be able to be lowered by following a healthy diet and including exercise in their daily routine. If diet and exercise don't lower blood pressure, medication may be needed. Adherence to the medication regimen is foremost in reducing blood pressure and preventing stroke.

For a patient who has had a stroke, start rehabilitation at admission. The amount of teaching you'll have to do depends on the extent of neurologic deficit.

- If necessary, teach the patient to comb his hair, dress, and wash. With the aid of a physical therapist and an occupational therapist, obtain appliances, such as walking frames, hand bars by the toilet, and ramps, as needed. The patient may fail to recognize that he has a paralyzed side (called unilateral neglect) and must be taught to inspect that side of his body for injury and to protect it from harm. If speech therapy is indicated, encourage the patient to begin as soon as possible and to follow through with the speech pathologist's suggestions. To reinforce teaching, involve the patient's family in all aspects of rehabilitation. With their cooperation and support, devise a realistic discharge plan,

  and let them help decide when the patient can return home.
- Before discharge, warn the patient or his family to report any premonitory signs of a stroke, such as severe headache, drowsiness, confusion, and dizziness. Emphasize the importance of regular follow-up visits.
- If aspirin has been prescribed to minimize the risk of embolic stroke, tell the patient to watch for possible GI bleeding. Make sure the patient and his family realize that acetaminophen isn't a substitute for aspirin.

To help prevent stroke:

- Stress the need to control diseases, such as diabetes or hypertension.
- Teach all patients (especially those at high risk) the importance of following a low-cholesterol, low-salt diet; watching their weight; increasing activity; avoiding smoking and prolonged bed rest; and minimizing stress. (See *Preventing stroke*, page 195.)
- Ensure that the patient understands that if symptoms develop, he should go to the emergency department immediately.

Meningitis

In meningitis, the brain and the spinal cord meninges become inflamed, usually as a result of a viral or, less commonly, a bacterial infection. Such inflammation may involve all three meningeal membranes—the dura mater, the arachnoid, and the pia mater. Viral meningitis is usually

mild and often clears on its own in 10 days or less. Bacterial meningitis requires prompt treatment with I.V. antibiotics.

Causes and incidence

Bacterial meningitis is almost always a complication of another bacterial infection—bacteremia (especially from pneumonia, empyema, osteomyelitis, or endocarditis), sinusitis, otitis media, encephalitis, myelitis, or brain abscess—usually caused by *Neisseria meningitidis*, *Haemophilus influenzae* (in children and young adults), or *Streptococcus pneumoniae* (in adults). In some cases, a virus is suspected. (See *Lymphocytic choriomeningitis*.) Meningitis may also follow skull fracture, a penetrating head wound, lumbar puncture, or ventricular shunting procedure. Aseptic meningitis may result from a virus or other organism. Sometimes, no causative organism can be found. Meningitis commonly begins as an inflammation of the pia-arachnoid, which may progress to congestion of adjacent tissues and destruction of some nerve cells.

Incidence of meningitis is high among Blacks and Native Americans. Male infants have a high incidence of gram-negative neonatal meningitis.

Complications

- Visual impairment
- Optic neuritis
- Cranial nerve palsies
- Personality changes
- Headache
- Paresis or paralysis
- Vasculitis
- Endocarditis

Signs and symptoms

The cardinal signs of meningitis are infection (fever, chills, and malaise) and increased intracranial pressure (ICP; headache, vomiting and, rarely, papilledema). Signs of meningeal irritation include nuchal rigidity,

positive Brudzinski's and Kernig's signs, exaggerated and symmetrical deep tendon reflexes, and opisthotonos (a spasm in which the back and extremities arch backward so that the body rests on the head and heels). (See *Two signs of meningitis*, pages 198 and 199.) Other manifestations of meningitis are irritability; sinus arrhythmias; photophobia, diplopia, and other visual problems; and delirium, deep stupor, and coma.

An infant may not show clinical signs of infection but may be fretful and refuse to eat. Such an infant may vomit often, leading to dehydration; this prevents a bulging fontanel and thus masks this important sign of increased ICP. As the illness progresses, twitching, seizures (in 30% of infants), or coma may develop. Most older children have the same symptoms as adults. In subacute meningitis, the onset may be insidious.

LYMPHOCYTIC CHORIOMENINGITIS

Lymphocytic choriomeningitis (LCM) is a mild, biphasic, febrile illness lasting about 2 weeks. Infection occurs through inhalation of the LCM virus or arenavirus from infectious aerosolized particles of the host (rodents such as mice or hamsters) or its excreta (urine, feces, or saliva) 1 to 3 weeks before the onset of symptoms. It can also result from contact with food contaminated with the virus or by contamination of mucous membranes, skin lesions, or cuts with infected body fluids. Handlers of infected animals or their excreta are at risk for this disease. Most cases occur in the northeast and eastern seaboard areas of the United States. LCM is more common during fall and winter.

The incubation period is 8 to 13 days after exposure. Early characteristics include fever, malaise, anorexia, weakness, muscle aches, retro-orbital headache, nausea, and vomiting. Sore throat, nonproductive cough, joint pain, chest pain, testicular pain, and parotid (salivary gland) pain may occur. Meningeal symptoms appear in 15

to 21 days, with signs and symptoms of meningitis (fever, increased headache, and stiff neck) or encephalitis (drowsiness, confusion, sensory disturbances, and motor abnormalities such as paralysis). Alopecia may also occur.

Complications include temporary or permanent neurologic damage, possible maternal transmission (pregnancy-related infection is associated with spontaneous abortion, congenital hydrocephalus, chorioretinitis, and mental retardation), myelitis, Guillain-Barrétype syndrome, orchitis or parotitis, myocarditis, psychosis, joint pain and arthritis, and prolonged convalescence with continuing dizziness, somnolence, and fatigue.

Diagnosis is made by detection of immunoglobulin M antibodies by enzyme-linked immunosorbent assay from serum or cerebrospinal fluid (CSF) (the preferred diagnostic test). Lumbarpuncture CSF is typically abnormal and reveals increased opening pressure, increased protein levels, and a lymphocytic pleocytosis, usually in the range of several hundred white blood cells. Treatment is generally supportive and includes bed rest, anti-inflammatory drugs, and analgesics. Ribavirin has been shown to be effective against LCM in vitro. Acute hydrocephalus may require surgical shunting to relieve increased intracranial pressure.

Prevention involves teaching rodent control measures, basic hygiene practices, use of a personal respirator, importance of adequate ventilation, and use of a liquid disinfectant, such as a diluted household bleach solution, to clean areas with rodent droppings.

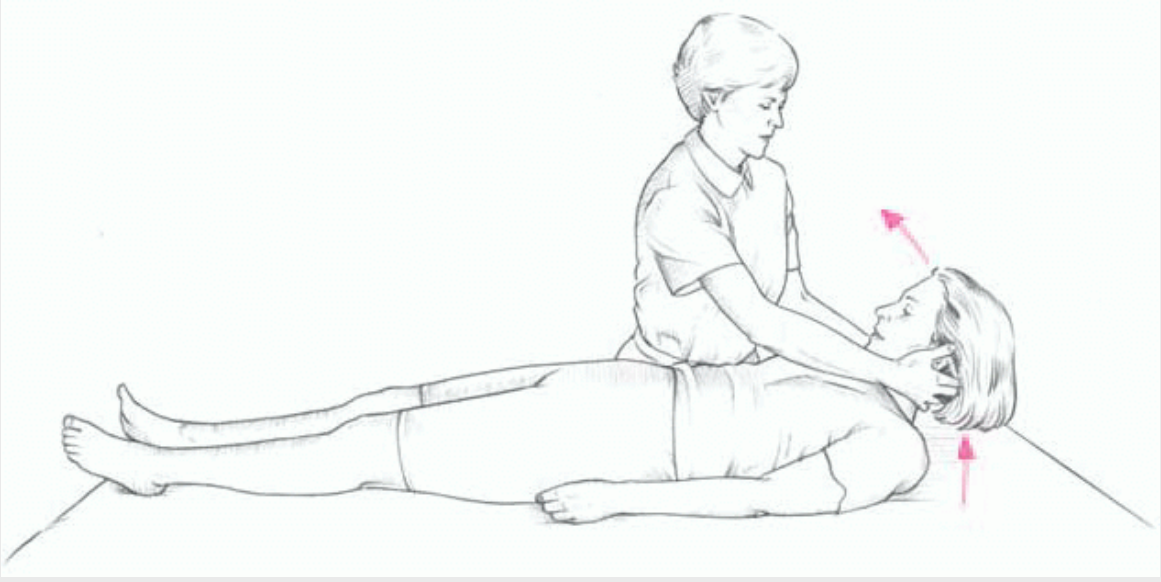
Diagnosis

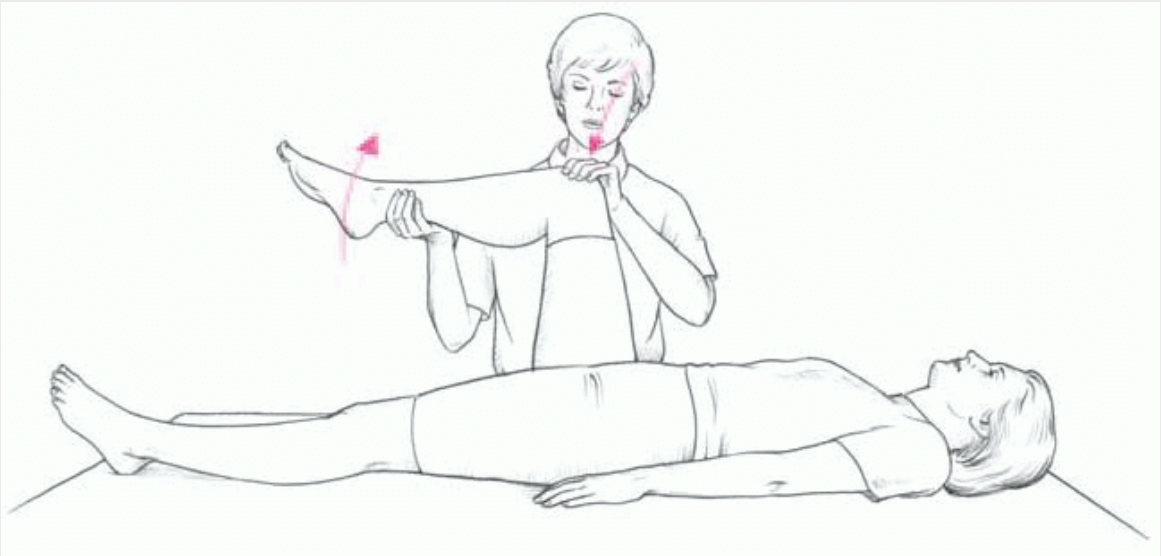
A lumbar puncture showing typical cerebrospinal fluid (CSF) findings, when accompanied by positive Brudzinski's and Kernig's signs, usually establishes a diagnosis. The following tests can uncover the primary sites of infection: cultures of blood, urine, and nose and throat secretions; chest X-ray; electrocardiogram; and a physical examination, with special attention to skin, ears, and sinuses. Lumbar puncture usually indicates elevated CSF pressure, from obstructed outflow at the arachnoid villi. The fluid may appear cloudy or milky white, depending on the number of white blood cells present. CSF protein levels tend to be high; glucose levels may be low. (In subacute meningitis, CSF findings may vary.) CSF culture and sensitivity tests usually identify the infecting organism, unless it's a virus. Leukocytosis and serum electrolyte abnormalities are also common. Computed

tomography scan can rule out cerebral hematoma, hemorrhage, or tumor.

TWO SIGNS OF MENINGITIS

Brudzinski's sign: Place the patient in a dorsal recumbent position, put your hands behind her neck, and bend it forward. Pain and resistance may indicate meningeal inflammation, neck injury, or arthritis. If the patient also flexes her hips and knees in response to this manipulation, chances are she has meningitis.





Kernig's sign: Place the patient in a supine position. Flex her leg at the hip and knee and then straighten the knee. Pain or resistance points to meningitis.

Treatment

Treatment of meningitis includes appropriate antibiotic therapy for bacterial meningitis and vigorous supportive care. Usually, I.V. antibiotics are given for at least 2 weeks, followed by oral antibiotics. Dexamethasone has been shown to be effective as adjunctive therapy in the treatment of meningitis caused by *H. influenzae* type B and in pneumococcal meningitis if given before the first dose of antibiotic. It has also been shown to reduce the incidence of deafness, a common complication of meningitis.

Other drugs include mannitol to decrease cerebral edema, an anticonvulsant (usually given I.V.) or sedative to reduce restlessness, and aspirin or acetaminophen to relieve headache and fever. Supportive measures include bed rest, fever reduction, and measures to prevent dehydration. The patient's room is kept darkened and quiet because any increase in sensory stimulation may cause a seizure. Isolation is necessary if nasal cultures are positive. Appropriate therapy for any coexisting conditions, such as endocarditis or pneumonia, is included as well. To prevent meningitis, prophylactic antibiotics are sometimes used

after ventricular shunting procedures, skull fracture, or penetrating head wounds, but this use is controversial.

For viral meningitis, treatment is supportive.

Special considerations

Patients must be watched carefully for changes in neurologic function or other signs of worsening condition.

- Assess neurologic function often. Observe level of consciousness (LOC) and signs of increased ICP (plucking at the bedcovers, vomiting, seizures, and a change in motor function and vital signs). Watch for signs of cranial nerve involvement (ptosis, strabismus, and diplopia).

ALERT

Be especially alert for a temperature increase up to 102° F (38.9° C), deteriorating LOC, onset of seizures, and altered respirations, all of which may signal an impending crisis.

- Monitor fluid balance. Maintain adequate fluid intake to avoid dehydration, but avoid fluid overload because of the danger of cerebral edema. Measure central venous pressure and intake and output accurately.
- Watch for adverse effects of I.V. antibiotics and other drugs. To avoid infiltration and phlebitis, check I.V. site often, and change the site according to hospital policy.
- Position the patient carefully to prevent joint stiffness and neck pain. Turn him often, according to a planned positioning schedule. Assist with range-of-motion exercises.
- Maintain adequate nutrition and elimination. It may be necessary to provide small, frequent meals or to supplement meals with nasogastric tube or parenteral feedings. To prevent constipation and minimize the risk of increased ICP resulting from straining at stool, give the patient a mild laxative or stool softener.

- Ensure the patient's comfort. Provide mouth care regularly. Maintain a quiet environment. Darkening the room may decrease photophobia. Relieve headache with a nonopioid analgesic, such as aspirin or acetaminophen as ordered. (Opioids interfere with accurate neurologic assessment.)
- Provide reassurance and support. The patient may be frightened by his illness and frequent lumbar punctures. If he's delirious or confused, attempt to reorient him often. Reassure his family that the delirium and behavior changes caused by meningitis usually disappear. However, if a severe neurologic deficit appears permanent, refer the patient to a rehabilitation program as soon as the acute phase of this illness has passed.
- To help prevent development of meningitis, teach patients with chronic sinusitis or other chronic infections the importance of proper medical treatment. Follow strict sterile technique when treating patients with head wounds or skull fractures.

PREVENTION

- *Give* Haemophilus influenzae *Type B and pneumococcal vaccines to children.*
- *Give meningococcal vaccine to college students.*
- *Give prophylactic antibiotics to those who have been exposed to a patient with meningitis.*

Encephalitis

Encephalitis is a severe inflammation of the brain, commonly caused by a mosquito-borne or, in some areas, a tick-borne virus. However, transmission by means other than arthropod bites may occur through ingestion of infected goat's milk and accidental injection or inhalation of the virus. Person-to-person, airborne transmission of viruses (such as measles or mumps) may also lead to encephalitis in nonimmunized populations. Eastern equine encephalitis may produce permanent neurologic damage and is commonly fatal. (See *Types of encephalitis*, pages 201 to 203.)

In encephalitis, intense lymphocytic infiltration of brain tissues and the leptomeninges causes cerebral edema, degeneration of the brain's ganglion cells, and diffuse nerve cell destruction.

Causes and incidence

Encephalitis typically results from infection with arboviruses specific to rural areas. However, in urban areas, it's most frequently caused by enteroviruses (coxsackievirus, poliovirus, and echovirus) and flaviviruses (West Nile virus). Other causes include herpesvirus, mumps virus, human immunodeficiency virus, adenoviruses, rabies, and demyelinating diseases after measles, varicella, rubella, or vaccination.

Complications

- Bronchial pneumonia
- Urine retention
- Urinary tract infection
- Pressure ulcers
- Seizure disorder
- Parkinsonism
- Mental deterioration
- Coma

Signs and symptoms

All viral forms of encephalitis have similar clinical features, although certain differences do occur. Usually, the acute illness begins with sudden onset of fever, headache, and vomiting and progresses to include signs and symptoms of meningeal irritation (stiff neck and back) and neuronal damage (drowsiness, coma, paralysis, seizures, ataxia, tremors, nausea, vomiting, and organic psychoses). After the acute phase of the illness, coma may persist for days or weeks.

The severity of arbovirus encephalitis may range from subclinical to rapidly fatal necrotizing disease. Herpes encephalitis also produces signs and symptoms that vary from subclinical to acute and commonly fatal

fulminating disease. Associated effects include disturbances of taste or smell.

Diagnosis

During an encephalitis epidemic, diagnosis is readily made on clinical findings and patient history. However, sporadic cases are difficult to distinguish from other febrile illnesses, such as gastroenteritis or meningitis. Diagnosis may be assisted by serologic assays, such as immunoglobulin (Ig) Mcapture ELISA (MAC-ELISA) and Ig ELISA. Early in infection, IgM antibody is more specific, whereas later, IgG is more reactive. Monoclonal antibody studies show promise in diagnosis. Polymerase chain reaction is also being investigated.

When possible, identification of the virus in cerebrospinal fluid (CSF) or blood confirms this diagnosis. The common viruses that also cause herpes, measles, and mumps are easier to identify than arboviruses. Both herpesviruses and arboviruses can be isolated by inoculating young mice with a specimen taken from the patient. In herpes encephalitis, serologic studies may show rising titers of complement-fixing antibodies.

In all forms of encephalitis, CSF pressure is elevated and despite inflammation, the fluid is usually clear. White blood cell and protein levels in CSF are slightly elevated, but the glucose level remains normal. An EEG reveals abnormalities. Occasionally, a computed tomography scan or magnetic resonance imaging may be ordered to rule out cerebral hematoma.

Treatment

The antiviral acyclovir must be prescribed for herpes encephalitis. Antibiotics may be prescribed if the infection is thought to be caused by bacteria. Treatment of all other

forms of encephalitis is entirely supportive. Drug therapy includes phenytoin or another anticonvulsant, usually given I.V.; steroids such as dexamethasone to reduce cerebral inflammation and edema; corticosteroids; mannitol to reduce cerebral swelling (although the evidence for benefit is weak); sedatives for restlessness; and aspirin or acetaminophen to relieve headache and reduce fever. Ribavirin and

interferon alpha-2b were found to have some effect on West Nile encephalitis. Other supportive measures include adequate fluid and electrolyte intake to prevent dehydration and antibiotics for an associated infection such as pneumonia. Isolation is unnecessary. (See *Preventing mosquito-borne encephalitis.*)

TYPES OF ENCEPHALITIS

Four main virus agents cause most cases of encephalitis in the United States: eastern equine encephalitis (EEE), western equine encephalitis (WEE), St. Louis encephalitis, and La Crosse (LAC) encephalitis, all of which are transmitted by mosquitoes. Another virus, Powassan (POW), is a minor cause of encephalitis in the northern United States; this virus is transmitted by ticks. Most cases of arboviral encephalitis occur from June through September, when arthropods are most active. In milder parts of the country, where arthropods are active late into the year, cases can occur into the winter months.

No vaccines are available for these U.S.-based diseases. However, a Japanese encephalitis (JE) vaccine is available for those who will be traveling to Japan, a tick-borne encephalitis vaccine is available for those who will be traveling to Europe, and an equine vaccine is available for EEE, WEE, and Venezuelan equine encephalitis (VEE). Public health measures often require spraying of insecticides to kill larvae and adult mosquitoes as well as controlling standing water that can provide mosquito breeding sites.

Eastern equine encephalitis

- EEE is caused by an alphavirus virus transmitted to humans and horses by the bite of an infected mosquito.
- Incubation is 4 to 10 days.

- Symptoms begin with a sudden onset of fever, general muscle pains, and a headache of increasing severity; can progress to seizures and coma.
- One-third of those afflicted will die from the disease and of those who recover, many will suffer irreversible brain damage requiring care.
- Human cases are usually preceded by outbreaks in horses.
- The virus occurs in natural cycles involving birds in swampy areas nearly every year during the warm months. The virus doesn't escape from these areas, however, and this mosquito doesn't usually bite humans or other mammals.

Western equine encephalitis

- The alphavirus WEE is the causative agent. The virus is closely related to the EEE and VEE viruses
- The enzootic cycle of WEE involves passerine birds, in which the infection is inapparent, and culicine mosquitoes, principally *Culex tarsalis*, a species associated with irrigated agriculture and stream drainages.
- Human WEE cases are usually first seen in June or July.
- Most WEE infections are asymptomatic or present as mild, nonspecific illness. Patients with clinically apparent illness usually have a sudden onset with fever, headache, nausea, vomiting, anorexia, and malaise, followed by altered mental status, weakness, and signs of meningeal irritation.
- Children, especially those younger than age 1, are affected more severely than adults and may be left with permanent sequelae, which are seen in 5% to 30% of young patients.

- Mortality is about 3%.

St. Louis encephalitis

- The leading cause of St. Louis encephalitis is flaviviral. St. Louis encephalitis is the most common mosquito-transmitted human pathogen in the United States.
- Mosquitoes become infected by feeding on birds infected with the St. Louis encephalitis virus. Infected mosquitoes then transmit virus to humans and animals during the feeding process. The virus grows both in the infected mosquito and the infected bird, but doesn't make either one sick.
- Less than 1% of St. Louis encephalitis viral infections are clinically apparent; the majority are undiagnosed.
- Illness ranges in severity from a simple febrile headache to meningoencephalitis, with an overall case-fatality ratio of 5% to 15%.

- The incubation period is 5 to 15 days.
- Mild infections present with fever and headache. More severe infection is marked by headache, high fever, neck stiffness, stupor, disorientation, coma, tremors, occasional convulsions (especially in infants), and spastic (but rarely flaccid) paralysis.
- The disease is generally milder in children than in adults, but in those children who do have disease, there's a high rate of encephalitis.
- Elderly people are at highest risk for severe disease and death.
- During the summer season, St. Louis encephalitis virus is maintained in a mosquito-bird-mosquito cycle, with

periodic amplification by peridomestic birds and *Culex* mosquitoes.

La Crosse encephalitis

- The LAC virus, a bunyavirus, is a zoonotic pathogen cycled between the daytime-biting tree hole mosquito, *Aedes triseriatus*, and vertebrate amplifier hosts (chipmunks, tree squirrels) in deciduous forest habitats. The virus is maintained over the winter by transmission in mosquito eggs. If the female mosquito is infected, she may lay eggs that carry the virus. Vector uses artificial containers (tires, buckets, and so forth) in addition to tree holes.
- LAC encephalitis initially presents as a nonspecific summertime illness with fever, headache, nausea, vomiting, and lethargy.
- Severe disease occurs most commonly in children younger than age 16 and is characterized by seizures, coma, paralysis, and a variety of neurological sequelae after recovery.
- Death occurs in less than 1% of clinical cases.
- Cases are often reported as aseptic meningitis or viral encephalitis of unknown etiology.
- During an average year, about 75 cases of LAC encephalitis are reported to the Centers for Disease Control and Prevention.

Powassan encephalitis

- The POW virus is a flavivirus.
- Recently a Powassan-like virus was isolated from the deer tick, *Ixodes scapularis*. The virus has been recovered from ticks (*Ixodes marxi* and *Dermacentor andersoni*) and from the tissues of an eastern spotted skunk.

- It's a rare cause of acute viral encephalitis.
- Patients who recover may have residual neurologic problems.

Venezuelan equine encephalitis

- Like EEE and WEE viruses, VEE is an alphavirus that causes encephalitis in horses and humans. VEE is a significant veterinary and public health problem in Central and South America.
- Infection of humans with the VEE virus is less severe than with EEE and WEE viruses, and fatalities are rare.
- Adults usually develop only an influenza-like illness; overt encephalitis is usually confined to children.
- Effective VEE virus vaccines are available for equines.

Japanese encephalitis

- JE virus, which is related to St. Louis encephalitis, is a flavivirus. It's widespread throughout Asia.
- Epidemics occur in late summer in temperate regions, but the infection is enzootic and occurs throughout the year in many tropical areas of Asia.
- The virus is maintained in a cycle involving culicine mosquitoes and waterbirds. It's transmitted to humans by Culex mosquitoes, primarily *Culex tritaeniorhynchus,* which breed in rice fields.
- Mosquitoes become infected by feeding on domestic pigs and wild birds infected with the JE virus. Infected mosquitoes then transmit the virus to humans and animals during the feeding process. The virus is amplified in domestic pigs and wild birds.
- The incubation period is 5 to 14 days.

- Mild infections occur without apparent symptoms other than fever with headache. More severe infection is marked by quick onset, headache, high fever, neck stiffness, stupor, disorientation, coma, tremors, occasional seizures (especially in infants), and spastic (but rarely flaccid) paralysis.
- The illness resolves in 5 to 7 days if there's no central nervous system involvement.
- The mortality is less than 10%, but is higher in children and can exceed 30%. Neurologic sequelae in patients who recover are reported in up to 30% of cases.
- A vaccine is currently available for human use in the United States for individuals who might be traveling to endemic countries.

Tick-borne encephalitis

- Tick-borne encephalitis (TBE) is caused by two closely related flaviviruses. The eastern subtype causes Russian spring-summer encephalitis (RSSE) and is transmitted by *Ixodes persulcatus,* whereas the western subtype is transmitted by *Ixodes ricinus* and causes Central European encephalitis (CEE).
- RSSE is the more severe infection, having a mortality of up to 25% in some outbreaks, whereas mortality in CEE seldom exceeds 5%.
- The incubation period is 7 to 14 days.
- Infection usually presents as a mild, influenza-type illness or as benign, aseptic meningitis, but may result in fatal meningoencephalitis.
- Fever is often biphasic, and there may be severe headache and neck rigidity, with transient paralysis of the limbs, shoulders or, less commonly, the respiratory

musculature. A few patients are left with residual paralysis.

- Although the great majority of TBE infections follow exposure to ticks, infection has occurred through the ingestion of infected cows' or goats' milk.
- An inactivated TBE vaccine is currently available in Europe and Russia.

West Nile encephalitis

- West Nile virus (WNV) is a flavivirus belonging to the Japanese encephalitis serocomplex that includes the closely related St. Louis encephalitis virus, Kunjin, and Murray Valley encephalitis viruses, as well as others.
- St. Louis encephalitis and WNE viruses are related.
- WNV can infect a wide range of vertebrates; in humans it usually produces either asymptomatic infection or mild febrile disease, but can cause severe and fatal infection in a small percentage of patients.
- The incubation period is thought to range from 3 to 14 days.
- Symptoms generally last 3 to 6 days.
- Like St. Louis encephalitis virus, WNV is transmitted principally by *Culex* species mosquitoes, but also can be transmitted by *Aedes*, *Anopheles*, and other species.
- The mild form of WNV infection has presented as a febrile illness of sudden onset often accompanied by malaise, anorexia, nausea, vomiting, eye pain, headache, myalgia, rash, and lymphadenopathy.
- A minority of patients with severe disease develop a maculopapular or morbilliform rash involving the neck, trunk, arms, or legs. Some patients experience severe muscle weakness and flaccid paralysis. Neurological presentations include ataxia, cranial nerve

abnormalities, myelitis, optic neuritis, polyradiculitis, and seizures. Although not observed in recent outbreaks, myocarditis, pancreatitis, and fulminant hepatitis have been described.

Murray Valley encephalitis

- MVE is endemic in New Guinea and in parts of Australia.
- It's related to the St. Louis encephalitis, WN, and JE viruses.
- Infections are common, and the small number of fatalities have mostly been in children.

PREVENTING MOSQUITO-BORNE ENCEPHALITIS

Help patients prevent mosquito-borne encephalitis by urging them to follow these steps:

- Wear long-sleeved shirts and long pants if going outside between dusk and dawn.
- A mosquito repellent with a 10% to 30% concentration of N, N-diethylmeta-toluamide (DEET) is recommended and should be applied to skin and clothing. Don't use DEET on the hands of young children or on infants under 2 months of age. Cover an infant's stroller or playpen with mosquito netting when outside.
- Refrain from unnecessary activity in places where mosquitoes are most prevalent.
- Repair holes in screens on doors and windows to prevent mosquitos from entering the home.
- Eliminate areas that attract mosquitoes, such as standing water in birdbaths, wheelbarrows, flower pots, old tires, and unused containers. These items are excellent breeding areas for mosquitoes.

Special considerations

During the acute phase of the illness:

- Assess neurologic function often. Observe level of consciousness and signs of increased intracranial pressure (ICP) (increasing restlessness, plucking at the bedcovers, vomiting, seizures, and changes in pupil size, motor function, and vital signs—such as rising blood pressure, widening pulse pressure, and slowly falling pulse). Watch for cranial nerve involvement (ptosis, strabismus, and diplopia), abnormal sleep patterns, and behavior changes.
- Maintain adequate fluid intake to prevent dehydration, but avoid fluid overload, which may increase cerebral edema. Measure and record intake and output accurately and assess daily weights.
- Carefully position the patient to prevent joint stiffness and neck pain, and turn him often. Assist with range-of-motion exercises.
- Maintain adequate nutrition. It may be necessary to give the patient small, frequent meals or to supplement meals with nasogastric tube or parenteral feedings.
- To prevent constipation and minimize the risk of increased ICP resulting from straining at stool, give a mild laxative or stool softener.
- Provide mouth care.
- Maintain a quiet environment. Darkening the room may decrease photophobia and headache. If the patient naps during the day and is restless at night, plan daytime activities to minimize napping and promote sleep at night.
- Provide emotional support and reassurance because the patient is apt to be frightened by the illness and frequent diagnostic tests.
- If the patient is delirious or confused, attempt to reorient him often. Providing a calendar or a clock in the patient's room may be helpful.
- Reassure the patient and his family that behavior changes caused by encephalitis usually disappear. If a neurologic deficit is severe and

appears permanent, refer the patient to a rehabilitation program as soon as the acute phase has passed.

Brain abscess

Brain abscess, also known as an *intracranial abscess,* is a free or encapsulated collection of pus usually in the temporal lobe, cerebellum, or frontal lobes. It can vary in size and may occur singly or in more than one location.

Untreated brain abscess is usually fatal; even with treatment, the prognosis is only fair, and about 30% of patients develop focal seizures. Multiple metastatic abscesses secondary to systemic or other infections have the poorest prognosis.

Causes and incidence

Brain abscess is usually secondary to some other infection, especially otitis media, sinusitis, dental abscess, and mastoiditis. Other causes include subdural empyema; bacterial endocarditis; human immunodeficiency virus infection; bacteremia; pulmonary or pleural infection; pelvic, abdominal, and skin infections; and cranial trauma, such as a penetrating head wound or compound skull fracture. Brain abscess also occurs in those with congenital heart disease and congenital blood vessel abnormalities of the lungs such as Osler-Weber-Rendu disease. These disorders carry a high risk of infection of the heart or lungs, which can then spread to the brain. Penetrating head trauma or bacteremia usually leads to staphylococcal infection; pulmonary disease, to streptococcal infection.

Brain abscess usually begins with localized inflammatory necrosis and edema, septic thrombosis of vessels, and suppurative encephalitis. This is followed by thick encapsulation of accumulated pus and adjacent meningeal infiltration by neutrophils, lymphocytes, and plasma cells.

Brain abscess has a relatively low incidence. It has a 10% mortality rate.

Signs and symptoms

Onset varies according to cause, but generally brain abscess produces clinical effects similar to those of a brain tumor. Early symptoms result

from increased intracranial pressure (ICP) and include constant intractable headache, worsened by straining; nausea; vomiting; and focal or generalized seizures. Typical later symptoms include ocular disturbances, such as nystagmus, decreased vision, and unequal pupil size.

Other features differ with the site of the abscess:

- *temporal lobe abscess:* auditory-receptive dysphasia, central facial weakness, and hemiparesis
- *cerebellar abscess:* dizziness, coarse nystagmus, gaze weakness on lesion side, tremor, and ataxia
- *frontal lobe abscess:* expressive dysphasia, hemiparesis with unilateral motor seizure, drowsiness, inattention, and mental function impairment.

Signs of infection, such as fever, pallor, and bradycardia, are absent until late stages unless they result from the predisposing condition. If the abscess is encapsulated, they may never appear. Depending on abscess size and location, level of consciousness (LOC) varies from drowsiness to deep stupor.

Diagnosis

A history of infection—especially of the middle ear, mastoid, nasal sinuses, heart, or lungs—or a history of congenital heart disease, along with a physical examination showing such characteristic clinical features as increased ICP, points to brain abscess. A computed tomography (CT) scan or magnetic resonance imaging (MRI) and, occasionally, arteriography (which highlights brain abscess by a halo) help locate the site. Blood culture will reveal any bacteria in the blood stream. Chest X-ray may reveal lung infection.

Examination of cerebrospinal fluid can help confirm infection, but lumbar puncture is risky because it can release the increased ICP and provoke cerebral herniation. A CT-guided stereotactic biopsy may be performed to drain and culture the abscess. Other tests include culture and sensitivity

of drainage to identify the causative organism, skull X-rays, and a

radioisotope scan.

Treatment

Management of patients with brain abscess has become increasingly challenging because of the proliferation of unusual bacterial, fungal, and parasitic infections, particularly in immunocompromised patients. Therapy consists of antibiotics to combat the underlying infection and surgical aspiration or drainage of the abscess. Surgical drainage or excision may be performed (CT scan or MRI can help determine the need for these procedures). Antimicrobials may be injected directly into the mass. Administration of antibiotics for at least 2 weeks before surgery can reduce the risk of spreading infection.

Other treatments during the acute phase are palliative and supportive and include mechanical ventilation, administration of I.V. fluids with diuretics (urea or mannitol), and glucocorticoids (dexamethasone) to combat increased ICP and cerebral edema. Anticonvulsants, such as phenytoin and phenobarbital, help prevent seizures.

Special considerations

The patient with an acute brain abscess requires intensive monitoring.

- Frequently assess neurologic status, especially LOC, speech, and sensorimotor and cranial nerve functions. Watch for signs of increased ICP (decreased LOC, vomiting, abnormal pupil response, and depressed respirations), which may lead to cerebral herniation with such signs as fixed and dilated pupils, widened pulse pressure, bradycardia or tachycardia, and absent respirations.
- Assess and record vital signs at least every hour.
- Monitor fluid intake and output carefully because fluid overload can contribute to cerebral edema.

If surgery is necessary, explain the procedure to the patient and answer his questions. After surgery:

- Continue frequent neurologic assessment. Monitor vital signs and intake and output.

- Watch for signs of meningitis (nuchal rigidity, headaches, chills, and sweats), an ever-present threat.
- Be sure to change a damp dressing often, using sterile technique and noting the amount of drainage. Never allow bandages to remain damp. To promote drainage and prevent reaccumulation of purulent material in the abscess, position the patient on the operative side.
- If the patient remains stuporous or comatose for an extended period, give meticulous skin care to prevent pressure ulcers, and position him to preserve function and prevent contractures.
- Encourage the patient to ambulate as soon as possible to prevent immobility and encourage independence.

PREVENTION

- *Stress the need for treatment of otitis media, mastoiditis, dental abscess, and other infections.*
- *Give prophylactic antibiotics after compound skull fracture or penetrating head wound.*

Huntington's disease

Also called *Huntington's chorea,* hereditary chorea, chronic progressive chorea, or adult chorea, Huntington's disease is a hereditary disease in which degeneration in the cerebral cortex and basal ganglia causes chronic progressive chorea and mental deterioration, ending in dementia.

Causes and incidence

Huntingdon's disease is inherited as a single faulty gene on chromosome #4 whereby part of the gene is repeated in multiple copies. It's transmitted as an autosomal dominant trait; either sex can transmit and inherit it. Each child of a parent with this disease has a 50% chance of inheriting it.

The disease usually strikes people between ages 35 and 55; however, 2% of cases occur in children, and 5% of cases occur as late as age 60. Death usually results 10 to 15 years after onset, from suicide, heart failure, or

pneumonia. Genetic testing is available for persons with a family history of the disease.

Complications

- Aspiration
- Pneumonia
- Heart failure
- Infections

Signs and symptoms

Onset is insidious. The patient eventually becomes totally dependent—emotionally and physically—through loss of musculoskeletal control, and he develops progressively severe choreic movements. Such movements are rapid, usually violent, and purposeless. Initially, they're unilateral and more prominent in the face and arms than in the legs, progressing from mild fidgeting to grimacing, tongue smacking, dysarthria (indistinct speech), athetoid movements (especially of the hands) related to emotional state, and torticollis.

Ultimately, the patient with Huntington's disease develops progressive dementia, although the dementia doesn't always progress at the same rate as the chorea. Dementia can be mild at first, but eventually causes severe disruption of the personality. Personality changes include obstinacy, carelessness, untidiness, moodiness, apathy, inappropriate behavior, loss of memory and concentration and, occasionally, paranoia.

Diagnosis

Huntington's disease can be detected by positron emission tomography and deoxyribonucleic acid analysis. Diagnosis is based on a characteristic clinical history: progressive chorea and dementia, onset in early middle age (35 to 40), and confirmation of a genetic link. Computed tomography scan and magnetic resonance imaging demonstrate brain atrophy. Molecular genetics may detect the gene for Huntington's disease in people at risk while they're still asymptomatic.

Treatment

Because Huntington's disease has no known cure, treatment is supportive, protective, and symptomatic. Dopamine blockers, such as phenothiazine or haloperidol, help control choreic movements and reduce abnormal behaviors. Reserpine and other drugs have been used with varying success with choreic movements and reduced abnormal behavior. Drugs, such as tetrabenazine and amantadine, are used to control extra movements. Some evidence suggests that co-enzyme Q10 may minimally decrease progression of the disease. Institutionalization may be necessary because of mental deterioration, which can't be halted or managed by drugs.

Special considerations

Patient comfort and support are the primary considerations.

- Provide physical support by attending to the patient's basic needs, such as hygiene, skin care, bowel and bladder care, and nutrition. Increase this support as mental and physical deterioration make him increasingly immobile.
- Offer emotional support to the patient and his family. Teach them about the disease, and listen to their concerns and special problems. Keep in mind the patient's dysarthria, and allow him extra time to express himself, thereby decreasing frustration. Teach the family to participate in the patient's care.
- Stay alert for possible suicide attempts. Control the patient's environment to protect him from suicide or other self-inflicted injury. Pad the side rails of the bed, but avoid restraints, which may cause the patient to injure himself with violent, uncontrolled movements.
- If the patient has difficulty walking, provide a walker to help him maintain his balance.
- Make sure affected families receive genetic counseling. All affected family members should realize that each of their offspring has a 50% chance of inheriting this disease.
- Refer people at risk who desire genetic testing to centers specializing in Huntington's care, where psychosocial support is available.

- Refer the patient and his family to the appropriate community organizations.
- For more information about this degenerative disease, refer the patient and his family to the Huntington's Disease Society of America.

Parkinson's disease

Named for James Parkinson, the English physician who wrote the first accurate description of the disease in 1817, Parkinson's disease characteristically produces progressive muscle rigidity, akinesia, involuntary tremor, and dementia. Death may result from aspiration pneumonia or an infection.

Causes and incidence

Although the cause of Parkinson's disease is unknown, study of the extrapyramidal brain nuclei (corpus striatum, globus pallidus, and substantia nigra) has established that a dopamine deficiency prevents affected brain cells from performing their normal inhibitory function within the central nervous system. Parkinson's disease occurs in families in some cases.

Parkinson's disease, also called *parkinsonism,* paralysis agitans, and shaking palsy, is one of the most common crippling diseases in the United States. Parkinson's disease strikes 2 in every 1,000 people, most often developing in those older than age 50; however, it also occurs in children and young adults. Because of increased longevity, this amounts to roughly 60,000 new cases diagnosed annually in the United States alone. Incidence increases in persons with repeated brain injury, including professional athletes, and persons using psychoactive substances, whether prescribed or illicit.

Complications

- Injury from falls
- Aspiration
- Urinary tract infections

- Skin breakdown

Signs and symptoms

The cardinal symptoms of Parkinson's disease are muscle rigidity and akinesia and an insidious resting tremor that begins in the fingers (unilateral pill-roll tremor), increases during stress or anxiety, and decreases with purposeful movement and sleep. Muscle rigidity results in resistance to passive muscle stretching, which may be uniform (lead-pipe rigidity) or jerky (cogwheel rigidity). Akinesia causes the patient to walk with difficulty (gait lacks normal parallel motion and may be retropulsive or propulsive) and produces a high-pitched, monotone voice; drooling; a masklike facial expression; loss of posture control (the patient walks with body bent forward); and dysarthria, dysphagia, or both. Occasionally, akinesia may also cause oculogyric crises (eyes are fixed upward, with involuntary tonic movements) or blepharospasm (eyelids are completely closed). Parkinson's disease itself doesn't impair the intellect, but a coexisting disorder, such as arteriosclerosis, may do so.

Diagnosis

Generally, laboratory data are of little value in identifying Parkinson's disease; consequently, diagnosis is based on the patient's age, history, and characteristic clinical picture.

Conclusive diagnosis is possible only after ruling out other causes of tremor, involutional depression, cerebral arteriosclerosis and, in patients younger than age 30, intracranial tumors, Wilson's disease, or phenothiazine or other drug toxicity.

Treatment

Because Parkinson's disease has no cure, the primary aim of treatment is to relieve symptoms and keep the patient functional as long as possible. Treatment consists of drugs, physical therapy and, in severe disease states unresponsive to drugs, stereotactic neurosurgery or the controversial treatment called fetal cell transplantation. In this treatment, fetal brain tissue is injected into the patient's brain. If the injected cells grow within the recipient's brain, they will allow the brain

to process dopamine, thereby either halting or reversing disease progression. Neurotransplantation techniques, including the use of nerve cells from other parts of the patient's body, have been attempted with varying results.

Drug therapy usually includes levodopa, a dopamine replacement that's most effective during early stages. It's given in increasing doses until symptoms are relieved or adverse effects appear. Because adverse effects can be serious, levodopa is usually given in combination with carbidopa to halt peripheral dopamine synthesis. Occasionally,

levodopa proves ineffective, producing dangerous adverse effects that include postural hypotension, hallucinations, and increased libido leading to inappropriate sexual behavior. In that case, alternative drug therapy includes anticholinergics such as trihexyphenidyl, antihistamines such as diphenhydramine, and amantadine, an antiviral agent.

Research on the oxidative stress theory has caused a controversy in drug therapy for Parkinson's disease. Traditionally, levodopa-carbidopa has been a first-line drug in management; however, it has also been associated with an acceleration of disease process. Inclusion of entacapone potentiates the effects of levodopa-carbidopa treatment so that less frequent doses are required.

Selegiline, an enzyme-inhibiting agent, allows conservation of dopamine and enhances the therapeutic effect of levodopa. Selegiline used with tocopherols delays the time when the patient with Parkinson's disease becomes disabled.

ELDER TIP

Elderly patients may need smaller doses of antiparkinsonian drugs because of reduced tolerance. Be alert for and report orthostatic hypotension, irregular pulse, blepharospasm, and anxiety or confusion.

When drug therapy fails, stereotactic neurosurgery, such as subthalamotomy and pallidotomy, may be an alternative. In these procedures, electrical coagulation, freezing, radioactivity, or ultrasound destroys the ventrolateral nucleus of the thalamus to prevent involuntary movement. This is most effective in young, otherwise

healthy people with unilateral tremor or muscle rigidity. Neurosurgery can only relieve symptoms. Brain stimulator implantation alters the activity of the area where Parkinson's disease symptoms originate. A pacemaker is implanted into the chest wall, and the electrode is threaded (using magnetic resonance imaging for guidance) to the thalamus, pallidum, or subthalamic nucleus. A successful procedure reduces the need for medication, thus reducing the medication-related adverse effects experienced by the patient.

Individually planned physical therapy complements drug treatment and neurosurgery to maintain normal muscle tone and function. Appropriate physical therapy includes both active and passive range-of-motion exercises, routine daily activities, walking, and baths and massage to help relax muscles.

Special considerations

Effectively caring for the patient with Parkinson's disease requires careful monitoring of drug treatment, emphasis on teaching self-reliance, and generous psychological support.

- Monitor drug treatment and adjust dosage, if necessary, to minimize adverse effects.
- If the patient has surgery, watch for signs of hemorrhage and increased intracranial pressure by frequently checking level of consciousness and vital signs.
- Encourage independence. The patient with excessive tremor may achieve partial control of his body by sitting on a chair and using its arms to steady himself. Advise the patient to change position slowly and dangle his legs before getting out of bed. Remember that fatigue may cause him to depend more on others.
- Help the patient overcome problems related to eating and elimination. For example, if he has difficulty eating, offer supplementary or small, frequent meals to increase caloric intake. Help establish a regular bowel routine by encouraging him to drink at least 2 qt (about 2 L) of liquids daily and eat high-fiber foods. He may need an elevated toilet seat to assist him from a standing to a sitting position.

- Give the patient and his family emotional support. Teach them about the disease, its progressive stages, and drug adverse effects. Show the family how to prevent pressure ulcers and contractures by proper positioning. Inform them of the dietary restrictions levodopa imposes, and explain household safety measures to prevent accidents. Help the patient and his family express their feelings and frustrations about the progressively debilitating effects of the disease. Establish long- and short-term treatment goals, and be aware of the patient's need for intellectual stimulation and diversion. Refer the patient and his family to the National Parkinson Foundation or

  the United Parkinson Foundation for more information.

Myelitis and acute transverse myelitis

Myelitis, or inflammation of the spinal cord, can result from several diseases. Poliomyelitis affects the cord's gray matter and produces motor dysfunction; leukomyelitis affects only the white matter and produces sensory dysfunction. These types of myelitis can attack any level of the spinal cord, causing partial destruction or scattered lesions. Acute transverse myelitis, which affects the entire thickness of the spinal cord, produces both motor and sensory dysfunctions. It has a rapid onset and is the most devastating form of myelitis.

The prognosis depends on the severity of cord damage and prevention of complications. If spinal cord necrosis occurs, prognosis for complete recovery is poor. Even without necrosis, residual neurologic deficits usually persist after recovery. Patients who develop spastic reflexes early in the course of the illness are more likely to recover than those who don't.

Causes and incidence

Acute transverse myelitis has a variety of causes. It commonly follows acute infectious diseases, such as measles or pneumonia (the inflammation occurs after the infection has subsided), and primary infections of the spinal cord itself, such as syphilis or acute disseminated encephalomyelitis. Acute transverse myelitis can accompany demyelinating diseases, such as an acute exacerbation of multiple

sclerosis, and inflammatory and necrotizing disorders of the spinal cord such as hematomyelia.

Certain toxic agents (carbon monoxide, lead, and arsenic) can cause a type of myelitis in which acute inflammation (followed by hemorrhage and possible necrosis) destroys the entire circumference (myelin, axis cylinders, and neurons) of the spinal cord. Other forms of myelitis may result from poliovirus, herpes zoster, herpesvirus B, or rabies virus; disorders that cause meningeal inflammation, such as syphilis, abscesses and other suppurative conditions, and tuberculosis; smallpox or polio vaccination; parasitic and fungal infections; and chronic adhesive arachnoiditis.

Peak incidence occurs between ages 10 and 19, then again between ages 30 and 39. Approximately 1,400 new cases are diagnosed each year in the United States. About 33,000 Americans have some type of disability from this disorder.

Complications

- Hypertension
- Urinary tract infection
- Urolithiasis
- Pneumonia
- Skeletal and smooth muscle deformities
- Myocarditis
- Paralytic ileus

Signs and symptoms

In acute transverse myelitis, onset is rapid, with motor and sensory dysfunctions below the level of spinal cord damage appearing in 1 to 2 days.

Patients with acute transverse myelitis develop flaccid paralysis of the legs (sometimes beginning in just one leg) with loss of sensory and sphincter functions. Such sensory loss may follow pain in the legs or trunk. Reflexes disappear in the early stages but may reappear later.

The extent of damage depends on the level of the spinal cord affected; transverse myelitis seldom involves the arms. If spinal cord damage is severe, it may cause shock (hypotension and hypothermia).

Diagnosis

Paraplegia of rapid onset usually points to acute transverse myelitis. In such patients, neurologic examination confirms paraplegia or neurologic deficit below the level of the spinal cord lesion and absent (or, in later stages) hyperactive reflexes. Cerebrospinal fluid may be normal or show increased lymphocytes or elevated protein levels. Diagnostic evaluation must rule out spinal cord tumor and identify the cause of any underlying infection.

Treatment

No effective treatment exists for acute transverse myelitis. However, this condition requires appropriate treatment of any

underlying infection. Some patients with postinfectious or multiple sclerosis-induced myelitis have received steroid therapy, but its benefits aren't clear.

Special considerations

Managing symptoms and treating the underlying infection are the primary considerations.

- Frequently assess vital signs. Watch carefully for signs of spinal shock (hypotension and excessive sweating).
- Prevent contractures with range-of-motion exercises and proper alignment.
- Watch for signs of urinary tract infections from indwelling urinary catheters.
- Prevent skin infections and pressure ulcers with meticulous skin care. Check pressure points often and keep skin clean and dry; use a low-pressure specialty or rotational bed or other pressure-relieving device.

- Initiate rehabilitation immediately. Assist the patient with physical therapy, bowel and bladder training, and the lifestyle changes his condition requires.

Alzheimer's disease

Alzheimer's disease, also called *primary degenerative dementia,* accounts for more than half of all dementias. It results in memory loss, confusion, impaired judgment, personality changes, disorientation, and loss of language skills. Because this is a primary progressive dementia, the prognosis for a patient with this disease is poor.

Causes and incidence

The cause of Alzheimer's disease is unknown; however, several factors are thought to be implicated in this disease. These include *neurochemical factors,* such as deficiencies in the neurotransmitter acetylcholine, somatostatin, substance P, and norepinephrine; *environmental factors;* and *genetic immunologic factors*. Genetic studies show that an autosomal dominant form of Alzheimer's disease is associated with early onset and early death, accounting for about 100,000 deaths a year. A family history of Alzheimer's disease and the presence of Down syndrome are two established risk factors. Alzheimer's disease isn't exclusive to the elder population; its onset begins in middle age in 1% to 10% of cases

The brain tissue of patients with Alzheimer's disease has three hallmark features: neurofibrillary tangles, neuritic plaques, and granulovascular degeneration. Examination of the brain after death also finds that it's atrophic, commonly weighing less than 1,000 g, compared with a normal brain weight of about 1,380 g.

About 360,000 new cases of Alzheimer's are diagnosed each year.

Complications

- Injury
- Pneumonia
- Infection

- Malnutrition
- Dehydration

Signs and symptoms

Onset is insidious. Initially, the patient undergoes almost imperceptible changes, such as forgetfulness, recent memory loss, difficulty learning and remembering new information, deterioration in personal hygiene and appearance, and an inability to concentrate. Gradually, tasks that require abstract thinking and activities that require judgment become more difficult. Progressive difficulty in communication and severe deterioration in memory, language, and motor function result in a loss of coordination and an inability to write or speak. Personality changes (restlessness, irritability) and nocturnal awakenings are common.

Patients also exhibit loss of eye contact, a fearful look, wringing of the hands, and other signs of anxiety. When a patient with Alzheimer's disease is overwhelmed with anxiety, he becomes dysfunctional, acutely confused, agitated, compulsive, or fearful.

Eventually, the patient becomes disoriented, and emotional lability and physical and intellectual disability progress. The patient becomes susceptible to infection and accidents. Usually, death results from infection.

ORGANIC BRAIN SYNDROME

Although many behavioral disturbances are clearly linked to organic brain dysfunction, the clinical syndromes associated with this type of impairment are sometimes hard to detect because they aren't always determined by the affected area of the brain or even by the extent of tissue damage. Instead, the way the patient's personality interacts with the brain injury determines the specific clinical effects. General symptoms often include impairment of orientation, memory, and intellectual and emotional function. These primary cognitive deficits help

to distinguish organic brain syndromes from neurosis and depression.

Diagnosis

Diagnosis of an organic brain syndrome depends on a detailed history of the onset of cognitive and behavioral disturbances; a complete neurologic assessment; and such tests as EEG, computed tomography scans, brain X-rays, cerebrospinal fluid analysis, and psychological studies. Organic brain syndromes are classified by etiology and specific clinical effects. Causes include infection, brain trauma, nutritional deficiency, cerebrovascular disease, degenerative disease, tumor, toxins, and metabolic or endocrine disorders.

Treatment

Effective treatment requires correction of the underlying cause. Special considerations may include reality orientation, emotional support for the patient and his family, a safe environment, mat therapy for an agitated or aggressive patient, and referral for psychological counseling.

Diagnosis

Early diagnosis of Alzheimer's disease is difficult because the patient's signs and symptoms are subtle. (See *Organic brain syndrome*.) Diagnosis relies on an accurate history from a reliable family member, mental status and neurologic examinations, and psychometric testing. A positron emission tomography scan measures the metabolic activity of the cerebral cortex and may help in early diagnosis. An EEG and a computed tomography scan may help in later diagnosis. Currently, the disease is diagnosed by exclusion; that is, tests are performed to rule out other disorders. The presence of Alzheimer's can't be confirmed until death, when pathologic findings are revealed at autopsy.

Treatment

Therapy consists of attempts to slow disease progression, manage behavioral problems, modify the home environment, and elicit family support. Some medications have proven helpful. Tacrine, a centrally acting anticholinesterase agent, is given to treat memory deficits. It has slowed progression of the disease and improved cognitive function in some patients. Other agents include donepezil and rivastigmine. Underlying disorders that contribute to the patient's confusion, such as hypoxia, are also identified and treated.

Special considerations

Overall care is focused on supporting the patient's remaining abilities and compensating for those lost.

- Establish an effective communication system with the patient and his family to help them adjust to the patient's altered cognitive abilities.
- Offer emotional support to the patient and his family members. Behavior problems may be worsened by excess stimulation or change in established routine. Teach them about the disease, and refer them to social service and community resources for legal and financial advice and support.
- Anxiety may cause the patient to become agitated or fearful. Intervene by helping him focus on another activity.
- Provide the patient with a safe environment. Encourage him to exercise, as ordered, to help maintain mobility.

UNDERSTANDING vCJD

Like conventional Creutzfeldt-Jakob disease (CJD), the variant of the disease (vCJD) is a rare, fatal neurodegenerative disease. Most cases have been reported in the United Kingdom. vCJD is most likely caused by exposure to bovine spongiform encephalopathy (BSE)—a fatal brain disease in cattle also known as *mad cow disease*—via ingestion of beef products from cattle with BSE.

vCJD affects patients at a much younger age (younger than age 55) than CJD, and the duration of the illness is much longer (14 months).

Regulations have been established in Europe to control outbreaks of BSE in cattle and to prevent contaminated meat from entering the food supply. The Centers for Disease Control and Prevention and the World Health Organization are still exploring vCJD and its relationship to BSE.

Creutzfeldt-Jakob disease

Creutzfeldt-Jakob disease (CJD) is a rare, rapidly progressive viral disease that attacks the central nervous system, causing dementia and neurologic signs and symptoms, such as myoclonic jerking, ataxia, aphasia, visual disturbances, and paralysis. CJD is always fatal. A new variant of CJD (vCJD) emerged in Europe in 1996. (See *Understanding vCJD*.)

Causes and incidence

The causative organism is difficult to identify because no foreign ribonucleic acid or deoxyribonucleic acid has been linked to the disease. CJD is believed to be caused by a specific protein called a prion, which lacks nucleic acids, resists proteolytic digestion, and spontaneously aggregates in the brain. Most cases are sporadic; 5% to 15% are familial, with an autosomal dominant pattern of inheritance. Although CJD isn't transmitted by normal casual contact, human-to-human transmission can occur as a result of certain medical procedures, such as corneal and cadaveric dura mater grafts. Isolated cases have resulted from childhood treatment with harvested human growth hormone and from improperly decontaminated neurosurgical instruments and brain electrodes.

CJD typically affects adults ages 40 to 65 and occurs in more than 50 countries. Males and females are affected equally. In people younger than age 30, incidence is 5 in 1 billion; in all other age groups, incidence is 1 in 1 million.

Complications

- Impaired gait
- Incontinence
- Akinetic mute state
- Bronchopneumonia
- Death

Signs and symptoms

Early signs and symptoms of mental impairment may include slowness in thinking, difficulty concentrating, impaired judgment, and memory loss. Dementia is progressive and occurs early. Involuntary movements, such as muscle twitching, trembling, and peculiar body movements, and visual disturbances, appear with disease progression and advancing mental deterioration. Hallucinations are also common. Duration of the typical illness is 4 months.

Diagnosis

CJD must be considered for anyone with signs of progressive dementia. Neurologic examination is the most effective tool in diagnosing CJD. Difficulty with rapid alternating movements and point-to-point movements are typically evident early in the disease.

An EEG may be performed to assess the patient for typical changes in brain wave activity. Computed tomography scan, magnetic resonance imaging of the brain, and lumbar puncture may be useful in ruling out other disorders that cause dementia. Though not diagnostic, presence of the 14-3-3 protein in the spinal fluid is highly suggestive of the disease when it's accompanied by other characteristic symptoms. Definitive diagnosis usually isn't obtained until an autopsy is done and brain tissue is examined.

Treatment

There's no cure for CJD, and its progress can't be slowed. Palliative care is provided to make the patient comfortable and to ease symptoms. Medications may be needed to control aggressive behaviors. These include sedatives and antipsychotics.

The need to provide a safe environment, control aggressive or agitated behavior, and meet physiologic needs may require monitoring and assistance in the home or in an institutionalized setting. Family counseling may help in coping with the changes required for home care.

Behavior modification may be helpful, in some cases, for controlling unacceptable or dangerous behaviors. Reality orientation, with repeated reinforcement of environmental and other cues, may help reduce disorientation.

Legal advice may be appropriate early in the course of the disorder regarding advance directives, power of attorney, and other legal actions that may make it easier to make ethical decisions regarding the care of an individual with CJD.

Special considerations

- Offer emotional support to the patient and his family. Teach them about the disease, and assist them through the grieving process. Refer the patient and his family to CJD support groups, and encourage participation.
- Contact social services and hospice, as appropriate, to assist the family with their needs.
- Encourage the patient and his family to discuss and complete advance directives.
- To prevent disease transmission, use caution when handling body fluids and other materials from patients suspected of having CJD.

Reye's syndrome

Reye's syndrome is an acute illness affecting children and, less commonly, adults. It causes fatty infiltration of the liver with concurrent hyperammonemia, encephalopathy, and increased intracranial pressure

(ICP). In addition, fatty infiltration of the kidneys, brain, and myocardium may occur.

Prognosis depends on the severity of central nervous system depression. Until recently, mortality was as high as 90%. Today, ICP monitoring and, consequently, early treatment of increased ICP, along with other treatment measures, have cut mortality to about 20%. Death is usually a result of cerebral edema or respiratory arrest. Comatose patients who survive may have residual brain damage.

Causes and incidence

Reye's syndrome typically begins within 1 to 3 days of an acute viral infection, such as an upper respiratory tract infection, type B influenza, or varicella (chickenpox). Incidence commonly rises during influenza outbreaks and may be linked to salicylate use. For this reason, use of aspirin for children younger than age 15 isn't recommended. The Reye's Syndrome Foundation warns against the use of salicylates, even in topical preparations, when a viral illness is suspected.

In Reye's syndrome, damaged hepatic mitochondria disrupt the urea cycle, which normally changes ammonia to urea for its excretion from the body. This results in hyperammonemia, hypoglycemia, and an increase in serum short-chain fatty acids, leading to encephalopathy. Simultaneously, fatty infiltration occurs in renal tubular cells, neuronal tissue, and muscle tissue, including the heart.

Reye's syndrome affects children from infancy to adolescence and occurs equally in boys and girls. Peak incidence is at age 6.

Complication

- Increased intracranial pressure

Signs and symptoms

The severity of the child's signs and symptoms varies with the degree of encephalopathy and cerebral edema. In any case,

Reye's syndrome develops in five stages. After the initial viral infection, a brief recovery period follows when the child doesn't seem seriously ill.

A few days later, he develops intractable vomiting; lethargy; rapidly changing mental status (mild to severe agitation, confusion, irritability, and delirium); rising blood pressure, respiratory rate, and pulse rate; and hyperactive reflexes.

Reye's syndrome commonly progresses to coma. As coma deepens, seizures develop, followed by decreased tendon reflexes and, usually, respiratory failure.

Increased ICP, a serious complication, is now considered the result of an increased cerebral blood volume causing intracranial hypertension. Such swelling may develop as a result of acidosis, increased cerebral metabolic rate, and an impaired autoregulatory mechanism.

Diagnosis

A history of a recent viral disorder with typical clinical features strongly suggests Reye's syndrome. An increased serum ammonia level, abnormal clotting studies, and hepatic dysfunction confirm it. Testing serum salicylate level rules out aspirin use. Absence of jaundice despite increased liver aminotransferase levels rules out acute hepatic failure and hepatic encephalopathy.

Abnormal test results may include:

- Liver function studies—aspartate aminotransferase and alanine aminotransferase elevated to twice normal levels; bilirubin level usually normal
- Liver biopsy—fatty droplets uniformly distributed throughout cells
- Cerebrospinal fluid (CSF) analysis— white blood cell count less than 10/mcl; with coma, increased CSF pressure.
- Coagulation studies—prothrombin time and partial thromboplastin time prolonged
- Blood values—serum ammonia levels elevated; serum glucose levels normal or, in 15% of cases, low; serum fatty acid and lactate levels increased.

STAGES OF TREATMENT FOR REYE'S SYNDROME

Signs and symptoms	Baseline treatment	Baseline intervention
Stage I: vomiting, lethargy, hepatic dysfunction	▪ To decrease intracranial pressure (ICP) and brain edema, give I.V. fluids at 2/3 maintenance. Also give an osmotic diuretic or furosemide. ▪ To treat hypoprothrombinemia, give vitamin K; if vitamin K is unsuccessful, give fresh frozen plasma. ▪ Monitor serum ammonia and blood glucose levels and plasma osmolality every 4 to 8 hours to check progress.	▪ Monitor the patient's vital signs and check his level of consciousness for increasing lethargy. Take vital signs more often as the patient's condition deteriorates. ▪ Monitor fluid intake and output to prevent fluid overload. Maintain urine output at 1.0 ml/kg/hour; plasma osmolality, 290 mOsm; and blood glucose, 150 mg/ml. (*Goal:* Keep glucose level high, osmolality normal to high, and ammonia level low.) Also, restrict protein.
Stage II: hyperventilation, delirium, hepatic dysfunction, hyperactive reflexes	▪ Continue baseline treatment.	▪ Maintain seizure precautions. ▪ Immediately report any signs of coma that require invasive, supportive therapy, such as intubation. ▪ Keep the head of the bed at a 30-degree angle.
Stage III: coma, hyperventilation, decorticate rigidity, hepatic dysfunction	▪ Continue baseline and seizure treatment. ▪ Monitor ICP with a subarachnoid screw or other invasive device. ▪ Provide endotracheal intubation and mechanical ventilation to control the partial pressure of arterial carbon dioxide ($PaCO_2$) levels. A paralyzing agent, such as atracurium or pancuronium	▪ Monitor ICP (should be less than 20 mm Hg before suctioning) or give a barbiturate I.V., as ordered; hyperventilate the patient as necessary. ▪ When ventilating the patient, maintain $PaCO_2$ between 25 and 30 mm Hg and the partial pressure of arterial oxygen between 80 and 100 mm Hg. ▪ Closely monitor cardiovascular status with a pulmonary artery

	I.V., may help maintain ventilation. ▪ Give mannitol I.V. or glycerol by nasogastric tube.	catheter or central venous pressure line. ▪ Give skin and mouth care and perform range-of-motion exercises.
Stage IV: deepening coma; decerebrate rigidity; large, fixed pupils; minimal hepatic dysfunction	▪ Continue baseline and supportive care. ▪ If all previous measures fail, some pediatric centers use barbiturate coma, decompressive craniotomy, hypothermia, or exchange transfusion.	▪ Check the patient for loss of reflexes and signs of flaccidity. ▪ Give the patent's family the extra support they need, considering their child's poor prognosis.
Stage V: seizures, loss of deep tendon reflexes, flaccidity, respiratory arrest, ammonia level above 300 mg/dl	▪ Continue baseline and supportive care.	▪ Help the patient's family to face his impending death.

Treatment

For treatment guidelines, see *Stages of treatment for Reye's syndrome*.

PREVENTION

Advise parents to give nonsalicylate analgesics and antipyretics such as acetaminophen. For more information, refer parents to the National Reye's Syndrome Foundation.

Guillain-Barré syndrome

Guillain-Barré syndrome is an acute, rapidly progressive, and potentially fatal form of polyneuritis that causes muscle weakness and mild distal sensory loss. Recovery is spontaneous and complete in about 95% of

patients, although mild motor or reflex deficits in the feet and legs may persist. The prognosis is best when symptoms clear between 15 and 20 days after onset.

Causes and incidence

Precisely what causes Guillain-Barré syndrome is unknown, but it may be a cell-mediated immunologic attack on peripheral nerves in response to a virus. The major pathologic effect is segmental demyelination of the peripheral nerves. Because this

syndrome causes inflammation and degenerative changes in both the posterior (sensory) and anterior (motor) nerve roots, signs of sensory and motor losses occur simultaneously.

This syndrome (also called infectious polyneuritis, Landry-Guillain-Barré syndrome, and acute idiopathic polyneuritis) can occur at any age but is most common between ages 30 and 50; it affects both sexes equally. In the United States, it has an incidence of 0.6 to 2.4 cases per 100,000 people.

Complications

- Thrombophlebitis
- Pressure ulcers
- Contractures
- Muscle wasting
- Aspiration
- Respiratory tract infections
- Respiratory and cardiac compromise

Signs and symptoms

About 50% of patients with Guillain-Barré syndrome have a history of minor febrile illness (10 to 14 days before onset), usually an upper respiratory tract infection or, less commonly, gastroenteritis with *Camphylobacter jejuni*. When infection precedes onset of Guillain-Barré

syndrome, signs of infection subside before neurologic features appear. Other possible precipitating factors include surgery, rabies or swine influenza vaccination, viral illness like Epstein-Barr virus, cytomegalovirus, hepatitis, and HIV, Hodgkin's or other malignant disease, and lupus erythematosus.

Symmetrical muscle weakness, the major neurologic sign, usually appears in the legs first (ascending type) and then extends to the arms and facial nerves in 24 to 72 hours. Sometimes, muscle weakness develops in the arms first (descending type) or in the arms and legs simultaneously. (See *Testing for thoracic sensation*.) In milder forms of this disease, muscle weakness may affect only the cranial nerves or may not occur at all.

Another common neurologic sign is paresthesia, which sometimes precedes muscle weakness but tends to vanish quickly. However, some patients with this

disorder never develop this symptom. Other clinical features may include facial diplegia (possibly with ophthalmoplegia), dysphagia or dysarthria and, less commonly, weakness of the muscles supplied by cranial nerve XI. Muscle weakness develops so quickly that muscle atrophy doesn't occur, but hypotonia and areflexia do. Stiffness and pain in the form of a severe “charley horse” commonly occur.

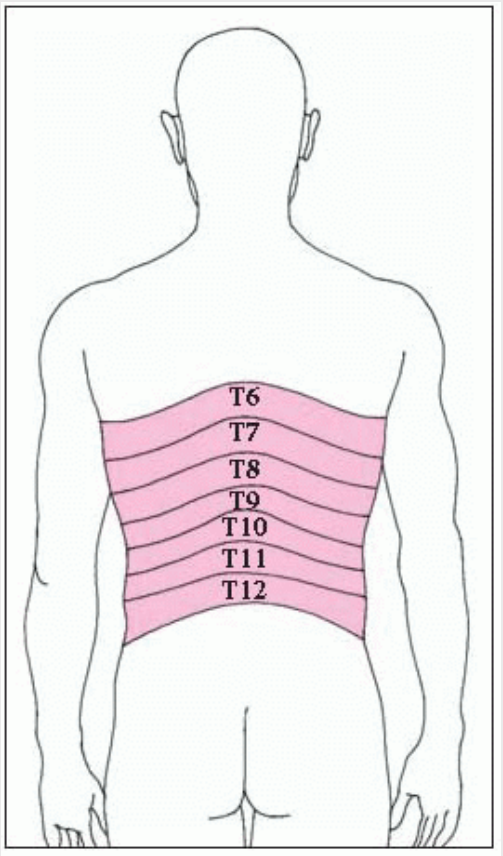
TESTING FOR THORACIC SENSATION

When Guillain-Barré syndrome progresses rapidly, test for ascending sensory loss by touching the patient or pressing his skin lightly with a pin every hour. Move systematically from the iliac crest (T12) to the scapula, occasionally substituting the blunt end of the pin to test the patient's ability to discriminate between sharp and dull.

Mark the level of diminished sensation to measure any change. If diminished sensation ascends to T8 or higher, the patient's intercostal muscle function (and consequently respiratory function) will probably be impaired. As Guillain-Barré syndrome subsides, sensory

and motor weakness descends to the lower thoracic segments, heralding a return of intercostal and extremity muscle function.

SEGMENTAL DISTRIBUTION OF SPINAL NERVES TO BACK OF THE BODY



Key: T = thoracic segments

The clinical course of Guillain-Barré syndrome is divided into three phases. The initial phase begins when the first definitive symptom appears and ends 1 to 3 weeks later, when no further deterioration manifests. The plateau phase lasts several days to 2 weeks and is followed by the recovery phase, which is believed to coincide with remyelination and axonal process regrowth. The recovery phase extends over a period of 4 to 6 months; patients with severe disease may take up to 2 years to recover, and recovery may not be complete.

Diagnosis

A history of febrile illness (usually a respiratory tract infection) and typical clinical features suggest Guillain-Barré syndrome.

Several days after onset of signs and symptoms, cerebrospinal fluid (CSF) protein levels begin to rise, peaking in 4 to 6 weeks, probably as a result of widespread inflammatory disease of the nerve roots. CSF white blood cell count remains normal, but in severe disease CSF pressure may rise above normal. Probably because of predisposing infection, CBC shows leukocytosis and a shift to immature forms early in the illness, but blood studies soon return to normal. Electromyography may show repeated firing of the same motor unit instead of widespread sectional stimulation. Nerve conduction velocities are slowed soon after paralysis develops and show demyelination. Diagnosis must rule out similar diseases such as acute poliomyelitis.

Treatment

Treatment is primarily supportive, including such measures as endotracheal (ET) intubation or tracheotomy if the patient has difficulty clearing secretions. Preventing complications is another goal of treatment.

Plasmapheresis is useful in decreasing severity of symptoms, thereby facilitating a more rapid recovery. I.V. immune globulin is equally effective in reducing the severity and duration of symptoms.

Special considerations

Monitoring the patient for escalation of symptoms is of special concern.

- Watch for ascending sensory loss, which precedes motor loss. Also, monitor vital signs and level of consciousness.
- Assess and treat respiratory dysfunction. If respiratory muscles are weak, take serial vital capacity recordings. Use a respirometer with a mouthpiece or a facemask for bedside testing.
- Obtain arterial blood gas measurements. Because neuromuscular disease results in primary hypoventilation with hypoxemia and hypercapnia, watch for respiratory failure. Be alert for signs of rising partial pressure of carbon dioxide (such as confusion and tachypnea).

- Auscultate breath sounds, turn and position the patient, and encourage coughing and deep breathing. Begin respiratory support at the first sign of dyspnea or with decreasing partial pressure of arterial oxygen.
- If respiratory failure becomes imminent, establish an emergency airway with an ET tube.
- Give meticulous skin care to prevent skin breakdown and contractures. Establish a strict turning schedule; inspect the skin (especially sacrum, heels, and ankles) for breakdown, and reposition the patient every 2 hours. After each position change, stimulate circulation by carefully massaging pressure points. Also, use foam, gel, or alternating pressure pads at points of contact.
- Perform passive range-of-motion exercises within the patient's pain limits. When the patient's condition stabilizes, change to gentle stretching and active assistance exercises.
- To prevent aspiration, test the gag reflex, and elevate the head of the bed before giving the patient anything to eat. If the gag reflex is absent, give nasogastric feedings until this reflex returns. If the patient has

  severe paralysis and is expected to have a long recovery period, a gastrostomy tube may be necessary to provide adequate nourishment.
- As the patient regains strength and can tolerate a vertical position, be alert for postural hypotension. Monitor blood pressure and pulse during tilting periods and, if necessary, apply toe-to-groin elastic bandages to prevent postural hypotension.
- Inspect the patient's legs regularly for signs of thrombophlebitis (localized pain, tenderness, erythema, edema, and positive Homans' sign), a common complication of Guillain-Barré syndrome. To prevent thrombophlebitis, apply antiembolism stockings and give prophylactic anticoagulants, as ordered.
- If the patient has facial paralysis, give eye and mouth care every 4 hours.
- Watch for urine retention. Measure and record intake and output every 8 hours, and offer the bedpan every 3 to 4 hours. Encourage

adequate fluid intake of 2 qt (about 2 L) per day, unless contraindicated. If urine retention develops, begin intermittent catheterization as ordered. Because the abdominal muscles are weak, the patient may need manual pressure on the bladder (Credé method) before he can urinate.

- To prevent or relieve constipation, offer the patient plenty of water, prune juice, and a high-bulk diet. If necessary, give daily or alternate-day suppositories (glycerin or bisacodyl) or enemas, as ordered.
- Before discharge, prepare a home care plan. Teach the patient how to transfer from bed to wheelchair, from wheelchair to toilet or tub, and how to walk short distances with a walker or a cane. Teach the family how to help him eat, compensating for facial weakness, and how to help him avoid skin breakdown. Stress the need for a regular bowel and bladder routine. Refer the patient for physical therapy as needed.
- Refer the patient's family to the Guillain-Barré Syndrome Foundation International.

NEUROMUSCULAR DISORDERS

Myasthenia gravis

Myasthenia gravis produces sporadic but progressive weakness and abnormal fatigability of striated (skeletal) muscles, exacerbated by exercise and repeated movement, but improved by anticholinesterase drugs. Usually, this disorder affects muscles innervated by the cranial nerves (face, lips, tongue, neck, and throat), but it can affect any muscle group. Myasthenia gravis follows an unpredictable course of recurring exacerbations and periodic remissions. There's no known cure. Drug treatment has improved prognosis and allows patients to lead relatively normal lives except during exacerbations. When the disease involves the respiratory system, it may be life threatening.

Causes and incidence

Myasthenia gravis causes a failure in transmission of nerve impulses at the neuromuscular junction. Theoretically, such impairment may result from an autoimmune response, ineffective acetylcholine release, or

inadequate muscle fiber response to acetylcholine. (See *Impaired transmission in myasthenia gravis*, page 220.)

Myasthenia gravis affects 3 of every 10,000 people at any age, but it's more common in young women and older men. About 20% of neonates born to mothers with myasthenia gravis have transient (or occasionally persistent) myasthenia. This disease may coexist with immunologic and thyroid disorders; about 15% of patients with myasthenia gravis have thymomas. Remissions occur in about 25% of patients.

Complications

- Respiratory distress
- Pneumonia
- Chewing and swallowing difficulties
- Aspiration

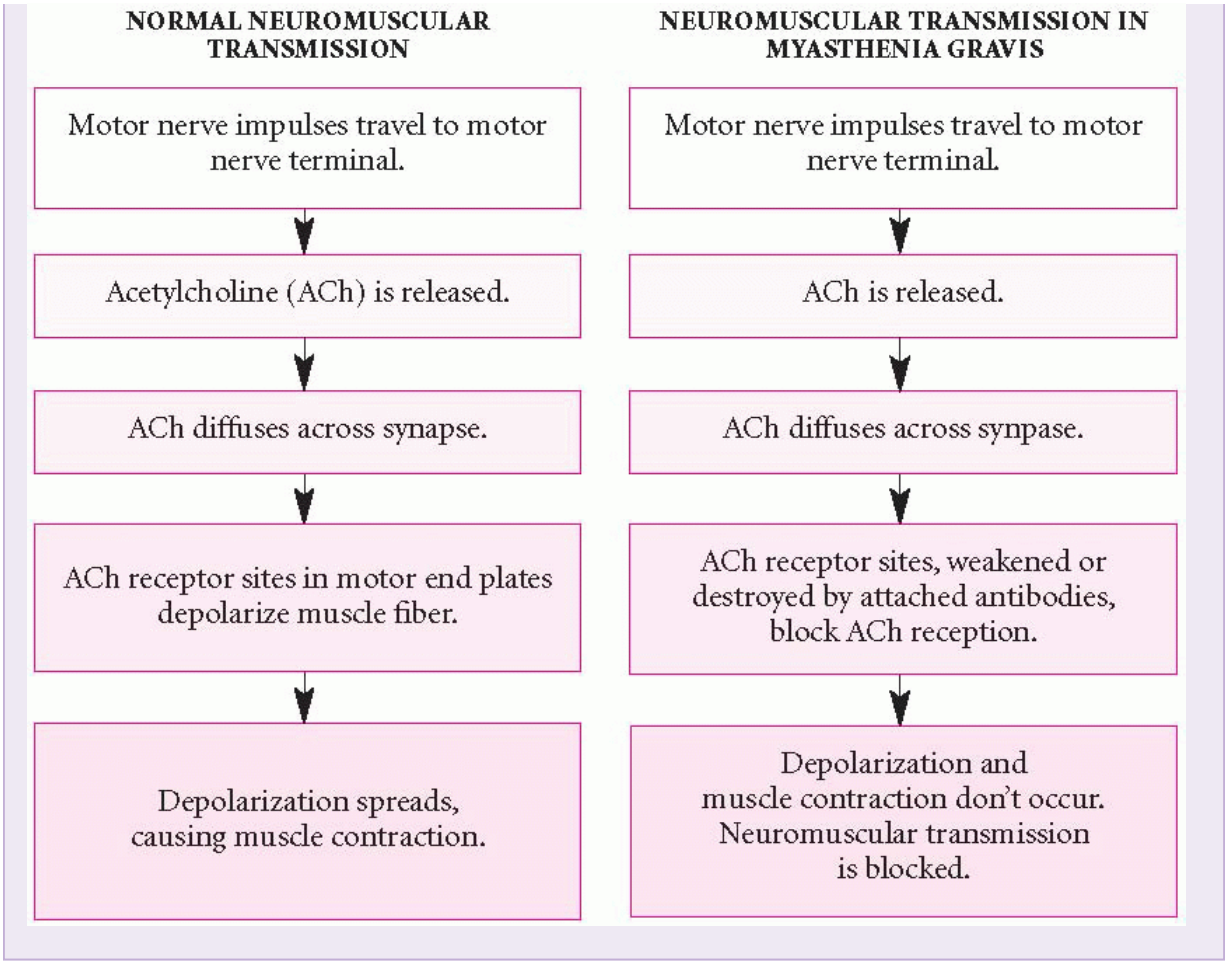
Signs and symptoms

The dominant symptoms of myasthenia gravis are skeletal muscle weakness and

fatigability. In the early stages, easy fatigability of certain muscles may appear with no other findings. Later, it may be severe enough to cause paralysis. Typically, myasthenic muscles are strongest in the morning but weaken throughout the day, especially after exercise. Short rest periods temporarily restore muscle function. Muscle weakness is progressive; more and more muscles become weak, and eventually some muscles may lose function entirely. Resulting symptoms depend on the muscle group affected; they become more intense during menses and after emotional stress, prolonged exposure to sunlight or cold, or infections.

PATHOPHYSIOLOGY

IMPAIRED TRANSMISSION IN MYASTHENIA GRAVIS



Onset may be sudden or insidious. In many patients, weak eye closure, ptosis, and diplopia are the first signs that something is wrong. Patients with myasthenia gravis usually have blank, expressionless faces and nasal vocal tones. They experience frequent nasal regurgitation of fluids and have difficulty chewing and swallowing. Because of this, they usually worry about choking. Their eyelids droop (ptosis), and they may have to tilt their heads back to see. Their neck muscles may become too weak to support their heads without bobbing.

In patients with weakened respiratory muscles, decreased tidal volume and vital capacity make breathing difficult and predispose to pneumonia and other respiratory tract infections. Respiratory muscle weakness (myasthenic crisis) may be severe enough to require an emergency airway and mechanical ventilation.

Diagnosis

Repeated muscle use over a very short time that fatigues and then improves with rest suggests a diagnosis of myasthenia gravis. Tests for this neurologic condition record

the effect of exercise and subsequent rest on muscle weakness. Electromyography, with repeated neural stimulation, may help confirm this diagnosis. Acetylcholine receptor antibodies may be present in the blood.

CONFIRMING DIAGNOSIS

The classic proof of myasthenia gravis is improved muscle function after an I.V. injection of edrophonium or neostigmine (anticholinesterase drugs).

In patients with myasthenia gravis, muscle function improves within 30 to 60 seconds and lasts up to 30 minutes. Longstanding ocular muscle dysfunction may fail to respond to such testing. This test can differentiate a myasthenic crisis from a cholinergic crisis (caused by acetylcholine overactivity at the neuromuscular junction). The acetylcholine receptor antibody titer may be elevated in generalized myasthenia. Evaluation should rule out thyroid disease and thymoma.

Treatment

Treatment is symptomatic. Anticholinesterase drugs, such as neostigmine and pyridostigmine, counteract fatigue and muscle weakness and allow about 80% of normal muscle function. However, these drugs become less effective as the disease worsens. Corticosteroids may relieve symptoms. Immunosuppressants are also used. Plasmapheresis is used in severe myasthenic exacerbation.

Patients with thymomas require thymectomy, which may cause remission in some cases of adult-onset myasthenia. Acute exacerbations that cause severe respiratory distress necessitate emergency treatment. Tracheotomy, positive-pressure ventilation, and vigorous suctioning to remove secretions usually produce improvement in a few days. Because anticholinesterase drugs aren't effective in myasthenic crisis, they're stopped until respiratory function improves. Myasthenic crisis requires immediate hospitalization and vigorous respiratory support.

Special considerations

Careful baseline assessment, early recognition and treatment of potential crises, supportive measures, and thorough patient teaching can minimize exacerbations and complications. Continuity of care is essential.

- Establish an accurate neurologic and respiratory baseline. Thereafter, monitor tidal volume and vital capacity regularly. The patient may need a ventilator and frequent suctioning to remove accumulating secretions.
- Be alert for signs of an impending crisis (increased muscle weakness, respiratory distress, and difficulty in talking or chewing).
- To prevent relapses, adhere closely to the ordered drug administration schedule. Be prepared to give atropine for anticholinesterase overdose or toxicity.
- Plan exercise, meals, patient care, and activities to make the most of energy peaks. For example, give medication 20 to 30 minutes before meals to facilitate chewing or swallowing. Allow the patient to participate in his care.
- When swallowing is difficult, give soft, solid foods instead of liquids to lessen the risk of choking.
- Patient teaching is essential because myasthenia gravis is usually a lifelong condition. Help the patient plan daily activities to coincide with energy peaks. Stress the need for frequent rest periods throughout the day. Emphasize that periodic remissions, exacerbations, and day-to-day fluctuations are common.
- Teach the patient how to recognize adverse effects and signs of toxicity of anticholinesterase drugs (headaches, weakness, sweating, abdominal cramps, nausea, vomiting, diarrhea, excessive salivation, and bronchospasm) and corticosteroids (euphoria, insomnia, edema, and increased appetite).
- Warn the patient to avoid strenuous exercise, stress, infection, and needless exposure to the sun or cold. All of these things may worsen signs and symptoms.

- For more information and an opportunity to meet other myasthenia gravis patients who lead full, productive lives, refer the patient to the Myasthenia Gravis Foundation.

Amyotrophic lateral sclerosis

Amyotrophic lateral sclerosis (ALS), also called *Lou Gehrig's disease,* is the most common of the motor neuron diseases that cause muscle atrophy. (See *Motor neuron disease*.) Other motor neuron diseases include progressive muscular atrophy and progressive bulbar palsy. Symptoms don't develop until age 50. A chronic, progressively debilitating disease, ALS may be fatal in less than 1 year or continue for 10 years or more, depending on the muscles it affects.

Causes and incidence

ALS affects about 1 of 100,000 people. The exact cause of ALS is unknown, but about 10% of cases have a genetic component. In these patients, it's an autosomal dominant trait and affects men and women equally.

Other than a family member affected with the hereditary form, there are no known risk factors.

Complications

- Pneumonia
- Aspiration
- Pressure ulcers
- Contractures

MOTOR NEURON DISEASE

In its final stages, motor neuron disease affects both upper and lower motor neuron cells. However, the site of initial cell damage varies according to the specific disease:

- *progressive bulbar palsy:* degeneration of upper motor neurons in the medulla oblongata
- *progressive muscular atrophy:* degeneration of lower motor neurons in the spinal cord
- *amyotrophic lateral sclerosis:* degeneration of upper motor neurons in the medulla oblongata and lower motor neurons in the spinal cord.

Signs and symptoms

Progressive loss of muscle strength and coordination eventually interfere with everyday activities. Patients with ALS develop fasciculations, accompanied by atrophy and weakness, especially in the muscles of the feet and the hands. Other signs include impaired speech; difficulty chewing, swallowing, and breathing and, occasionally, choking and excessive drooling. Mental deterioration doesn't occur, but patients may become depressed as a reaction to the disease.

Diagnosis

Characteristic clinical features indicate a combination of upper and lower motor neuron involvement without sensory impairment. Electromyography and muscle biopsy indicate that the motor nerves aren't functioning, yet sensory nerves are normal. Computed tomography scan and magnetic resonance imaging may help rule out other conditions, such as multiple sclerosis, spinal cord neoplasm, central nervous system syphilis, polyarteritis, syringomyelia, myasthenia gravis, progressive muscular dystrophy, and progressive strokes.

Treatment

Management aims to control symptoms and provide emotional, psychological, and physical support. Riluzole may increase quality of life and survival but doesn't reverse or stop disease progression.

Baclofen or diazepam help control spasticity that interferes with activities of daily living. Trihexyphenidyl or amitriptyline may be used for impaired ability to swallow saliva. Gastrostomy may be needed early

to prevent choking; referral to an otolaryngologist is advised. Physical therapy, rehabilitation, and use of appliances or orthopedic intervention may be required to maximize function. Devices to assist in breathing at night or mechanical ventilation should be discussed, but the patient's wishes should be respected.

Special considerations

Because mental status remains intact while progressive physical degeneration takes place, the patient acutely perceives every change. This threatens the patient's relationships, career, income, muscle coordination, sexuality, and energy.

- Implement a rehabilitation program designed to maintain independence as long as possible.
- Help the patient obtain assistive equipment, such as a walker and a wheelchair. Arrange for a visiting nurse to oversee the patient's status, to provide support, and to teach the family about the illness.
- Depending on the patient's muscular capacity, assist with bathing, personal hygiene, and transfers from wheelchair to bed. Help establish a regular bowel and bladder routine.
- To help the patient handle increased accumulation of secretions and dysphagia, teach him to suction himself. He should have a suction machine handy at home to reduce the fear of choking.
- To prevent skin breakdown, provide good skin care when the patient is bedridden. Turn him often, keep his skin clean and dry, and use pressure-reducing devices such as alternating air mattress.
- If the patient has trouble swallowing, give him soft, solid foods and position him upright during meals. Gastrostomy and nasogastric tube feedings may be necessary if he can no longer swallow. Teach the patient (if he's still able to feed himself) or his family how to administer gastrostomy feedings.
- Provide emotional support. A discussion of directives regarding health care decisions should be instituted before the patient becomes unable to communicate his wishes. Prepare the patient and his family

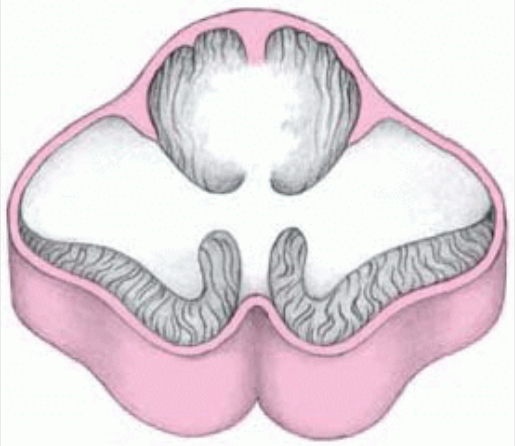
members for his eventual death, and encourage the start of the grieving process. Patients with ALS may benefit from a hospice program or the local ALS support group chapter.

Multiple sclerosis

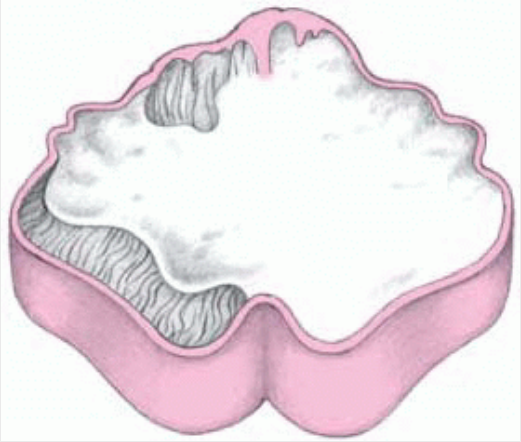
Multiple sclerosis (MS) is a progressive disease caused by demyelination of the white matter of the brain and spinal cord. (See *Demyelination in multiple sclerosis*.) In this disease, sporadic patches of demyelination throughout the central nervous system induce widely disseminated and varied neurologic dysfunction. Characterized by exacerbations and remissions, MS is a major cause of chronic disability in young adults.

DEMYELINATION IN MULTIPLE SCLEROSIS

Transverse section of cervical spine shows partial loss of myelin, characteristic of multiple sclerosis (MS). This degenerative process is called *demyelination*.



In this illustration, the loss of myelin is nearly complete. Clinical features of MS depend on the extent of demyelination.



The prognosis varies; MS may progress rapidly, disabling some patients by early adulthood or causing death within months of onset. However, 70% of patients lead active, productive lives with prolonged remissions.

Causes and incidence

The exact cause of MS is unknown, but current theories suggest a slow-acting or latent viral infection and an autoimmune response. Other theories suggest that environmental and genetic factors may also be linked to MS. Emotional stress, overwork, fatigue, pregnancy, and acute respiratory tract infections have been known to precede the onset of this illness.

MS usually begins between ages 20 and 40. It affects more women than men. A family history of MS and living in a geographical area with higher incidence of MS (northern Europe, northern United States, southern Australia, and New Zealand) increase the risk.

Complications

- Injuries from falls
- Urinary tract infections
- Constipation
- Joint contractures
- Pressure ulcers

- Rectal distention
- Pneumonia

Signs and symptoms

Clinical findings in MS depend on the extent and site of myelin destruction, the extent of remyelination, and the adequacy of subsequent restored synaptic transmission.

Signs and symptoms in MS may be transient, or they may last for hours or weeks. They may wax and wane with no predictable pattern, vary from day to day, and be bizarre and difficult for the patient to describe.

In most patients, visual problems and sensory impairment, such as numbness and tingling sensations (paresthesia), are the first signs that something may be wrong.

Other characteristic changes include:

- *ocular disturbances*—optic neuritis, diplopia, ophthalmoplegia, blurred vision, and nystagmus
- *muscle dysfunction*—weakness, paralysis ranging from monoplegia to quadriplegia, spasticity, hyperreflexia, intention tremor, and gait ataxia
- *urinary disturbances*—incontinence, frequency, urgency, and frequent infections
- *emotional lability*—characteristic mood swings, irritability, euphoria, and depression.

Associated signs and symptoms include poorly articulated or scanning speech and dysphagia. Clinical effects may be so mild that the patient is unaware of them or so bizarre that he appears hysterical.

Diagnosis

A misdiagnosis of psychiatric problems is common. Because early symptoms may be mild, years may elapse between onset of the first signs and the diagnosis, which typically requires evidence of multiple neurologic attacks and characteristic remissions and exacerbations. Magnetic resonance imaging may detect MS lesions; however, diagnosis

still remains difficult. Periodic testing and close observation of the patient are necessary, perhaps for years, depending on the course of the disease.

Abnormal EEG findings occur in one-third of patients. Lumbar puncture shows elevated gamma globulin fraction of immunoglobulin G but normal total cerebrospinal fluid (CSF) protein levels. Elevated CSF gamma globulin is significant only when serum gamma globulin levels are normal because it reflects hyperactivity of the immune system due to chronic demyelination. Oligoclonal bands of immunoglobulin can be detected when gamma globulin in CSF is examined by electrophoresis, and these bands are present in most patients, even when the percentage of gamma globulin in CSF is normal. In addition, the white blood cell count in CSF may rise. Differential diagnosis must rule out spinal cord compression, foramen magnum tumor (may mimic the exacerbations and remissions of MS), multiple small strokes, syphilis or other infection, and psychological disturbances.

Treatment

The aim of treatment is to shorten exacerbations and relieve neurologic deficits so that the patient can resume a normal lifestyle. Those with relapsing-remitting courses are placed on immune modulating therapy, with interferon or glatiramer acetate. Steroids are used to reduce the associated

edema of the myelin sheath during exacerbations.

Other drugs include baclofen, tizanidine, or diazepam to relieve spasticity, cholinergic agents to relieve urine retention and minimize frequency and urgency, amantadine to relieve fatigue, and antidepressants to help with mood or behavioral symptoms. During acute exacerbations, supportive measures include bed rest, comfort measures such as massages, prevention of fatigue, prevention of pressure ulcers, bowel and bladder training (if necessary), administration of antibiotics for bladder infections, physical therapy, and counseling. Physical therapy, speech therapy, occupational therapy, and support groups are also useful. Planned exercise programs help with maintaining muscle tone.

Special considerations

Management considerations focus on educating the patient and his family.

- Assist with physical therapy. Increase patient comfort with massages and relaxing baths. Assist with active, resistive, and stretching exercises to maintain muscle tone and joint mobility, decrease spasticity, improve coordination, and boost morale.
- Educate the patient and his family concerning the chronic course of MS. Emphasize the need to avoid temperature extremes, stress, fatigue, and infections and other illnesses, all of which can trigger an MS attack. Advise him to maintain independence by developing new ways of performing daily activities.
- Stress the importance of eating a nutritious, well-balanced diet that contains sufficient roughage and adequate fluids to prevent constipation.
- Evaluate the need for bowel and bladder training during hospitalization. Encourage adequate fluid intake and regular urination. Eventually, the patient may require urinary drainage by self-catheterization or, in men, condom drainage. Teach the correct use of suppositories to help establish a regular bowel schedule.
- Promote emotional stability. Help the patient establish a daily routine to maintain optimal functioning. Activity level is regulated by tolerance level. Encourage regular rest periods to prevent fatigue and daily physical exercise.
- Inform the patient that exacerbations are unpredictable, necessitating physical and emotional adjustments in lifestyle.
- For more information, refer him to the National Multiple Sclerosis Society.

CRANIAL NERVE DISORDERS

Trigeminal neuralgia

Trigeminal neuralgia, also called *tic douloureux,* is a painful disorder of one or more branches of the fifth cranial (trigeminal) nerve that produces paroxysmal attacks of excruciating facial pain precipitated by stimulation of a trigger zone. It can subside spontaneously, and remissions may last from several months to years.

Causes and incidence

Although the cause remains undetermined, trigeminal neuralgia may reflect an afferent reflex in the brain stem or in the sensory root of the trigeminal nerve. Such neuralgia may also be related to compression of the nerve root by posterior fossa tumors, middle fossa tumors, or vascular lesions (subclinical aneurysm), although such lesions usually produce simultaneous loss of sensation. Occasionally, trigeminal neuralgia is a manifestation of multiple sclerosis or herpes zoster. Whatever the cause, the pain of trigeminal neuralgia is probably produced by an interaction or shortcircuiting of touch and pain fibers.

Trigeminal neuralgia occurs mostly in people older than age 40, in women more commonly than men, and on the right side of the face more commonly than the left. Incidence is 4 to 5 cases per 100,000 people.

Signs and symptoms

Typically, the patient reports a searing or burning pain that occurs in lightninglike jabs and lasts from 1 to 15 minutes (usually 1 to 2 minutes) in an area innervated by one of the divisions of the trigeminal nerve, primarily the superior mandibular or maxillary division. The pain rarely affects more than one division and seldom the first division (ophthalmic) or both sides of the face. It affects the second (maxillary) and third (mandibular) divisions of the trigeminal nerve equally. (See *Trigeminal nerve function and distribution.*)

TRIGEMINAL NERVE FUNCTION AND DISTRIBUTION

Function

- Motor: chewing movements

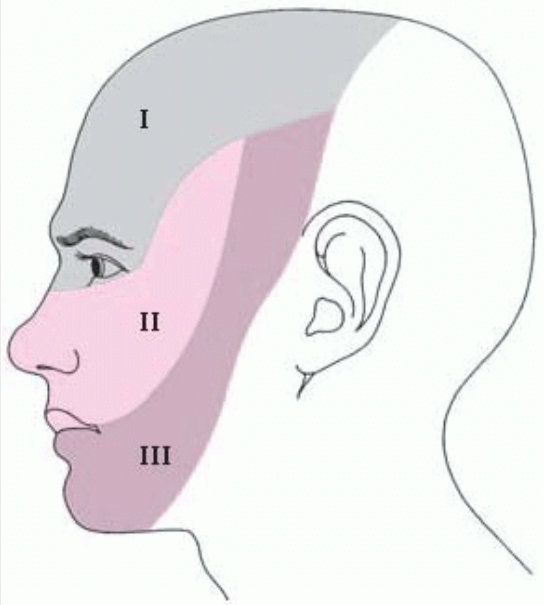
- Sensory: sensations of face, scalp, and teeth (mouth and nasal chamber)

Distribution

I Ophthalmic

II Maxillary

III Mandibular



These attacks characteristically follow stimulation of a trigger zone, usually by a light touch to a hypersensitive area, such as the tip of the nose, the cheeks, or the gums. Although attacks can occur at any time, they may follow a draft of air, exposure to heat or cold, eating, smiling, talking, or drinking hot or cold beverages. The frequency of attacks varies greatly, from many times a day to several times a month or year. Between attacks, most patients are free from pain, although some have a constant, dull ache. No patient is ever free from the fear of the next attack.

Diagnosis

The patient's pain history is the basis for diagnosis because trigeminal neuralgia produces no objective clinical or pathologic changes. Physical examination shows no impairment of sensory or motor function; indeed,

sensory impairment implies a space-occupying lesion as the cause of pain.

Observation during the examination shows the patient favoring (splinting) the affected area. To ward off a painful attack, the patient commonly holds his face immobile when talking. He may also leave the affected side of his face unwashed and unshaven or protect it with a coat or shawl. When asked where the pain occurs, he points to—but never touches—the affected area. Witnessing a typical attack helps to confirm diagnosis. Rarely, a tumor in the posterior fossa can produce pain that's clinically indistinguishable from trigeminal neuralgia. Skull X-rays, computed tomography scan, and magnetic resonance imaging rule out sinus or tooth infections and tumors. If the patient has trigeminal neuralgia, these test results are normal.

Treatment

Oral administration of carbamazepine (Tegretol), gabapentin (Neurontin), or phenytoin may temporarily relieve or prevent pain. Opioids are of little help during the pain episode. Caution should be used when treating a chronic problem with opioids.

When these medical measures fail or attacks become increasingly frequent or severe, neurosurgical procedures may provide permanent relief. The preferred procedure is percutaneous electrocoagulation of nerve rootlets under local anesthetic. New treatments include a percutaneous radio frequency procedure, which causes partial root destruction and relieves pain, and microsurgery for vascular decompression of the trigeminal nerve.

Special considerations

The focus here is management of pain.

- Observe and record the characteristics of each attack, including the patient's protective mechanisms.
- Provide adequate nutrition in small, frequent meals served at room temperature.
- Avoid jarring the bed and causing increased discomfort.

- If the patient is receiving neuroleptics, watch for cutaneous and hematologic reactions (erythematous and pruritic rashes, urticaria, photosensitivity, exfoliative dermatitis, leukopenia, agranulocytosis, eosinophilia, aplastic anemia, and thrombocytopenia) and, possibly, urine retention and transient drowsiness. For the first 3 months of therapy, monitor complete blood count and liver function weekly, then monthly thereafter. Warn the patient to immediately report fever, sore throat, mouth ulcers, easy bruising, or petechial or purpuric hemorrhage because these may signal thrombocytopenia or aplastic anemia and may require discontinuation of drug therapy.
- If the patient is receiving phenytoin, also watch for adverse effects, including ataxia, skin eruptions, gingival hyperplasia, and nystagmus.
- After resection of the first branch of the trigeminal nerve, tell the patient to avoid rubbing his eyes and using aerosol spray. Advise him to wear glasses or goggles outdoors and to blink often.
- After surgery to sever the second or third branch, tell the patient to avoid hot foods and drinks, which could burn his mouth, and to chew carefully to avoid biting his mouth. The patient may need to eat pureed food, possibly through a straw. Advise him to place food in the unaffected side of his mouth when chewing, to brush his teeth and rinse his mouth often, and to see the dentist twice a year to detect cavities because he won't experience pain from cavities in the area of the severed nerve.
- After surgical decompression of the root or partial nerve dissection, check neurologic and vital signs often.
- Provide emotional support, and encourage the patient to express his feelings. Promote independence through self-care and maximum physical activity. Reinforce natural avoidance of stimulation (air, heat, and cold) of trigger zones (lips, cheeks, and gums).

Bell's palsy

Bell's palsy is a disease of the seventh cranial nerve (facial) that produces unilateral or bilateral facial weakness or paralysis. (See *Recognizing unilateral Bell's palsy*, page 228.) Onset is rapid. In 80% to 90% of patients, Bell's palsy subsides spontaneously, with complete

recovery in 1 to 8 weeks; however, recovery may be delayed in the elderly. If recovery is partial, contractures may develop on the paralyzed side of the face. Bell's palsy may recur on the same or opposite side of the face.

Causes and incidence

Bell's palsy blocks the seventh cranial nerve, which is responsible for motor innervation of the muscles of the face. The conduction block is from an inflammatory reaction around the nerve (usually at the internal auditory meatus), which may result from infection, hemorrhage, tumor, meningitis, local trauma, hypertension, sarcoidosis, Lyme disease, or infarction of the nerve.

Bell's palsy affects all age groups and males and females nearly equally, although females are slightly more likely to develop it during their late teens and early 20s. In the United States, incidence is 23 cases per 100,000 people.

Complications

- Corneal ulcers
- Blindness
- Impaired nutrition

Signs and symptoms

Bell's palsy usually produces unilateral facial weakness, occasionally with aching pain around the angle of the jaw or behind

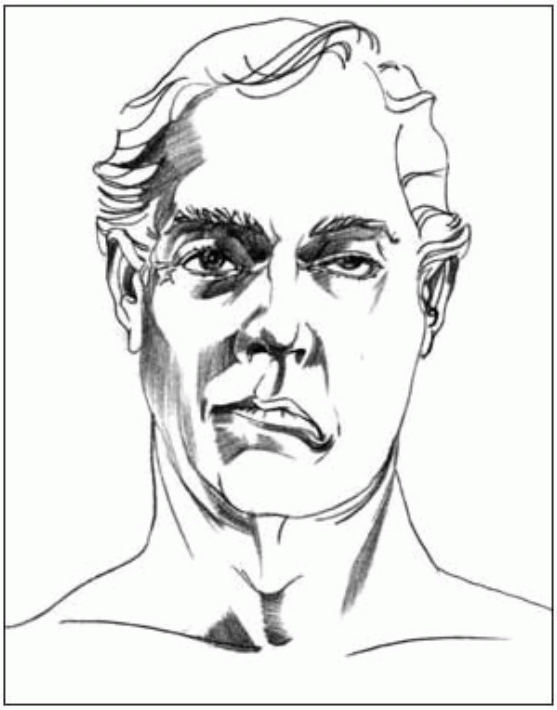
the ear. On the weak side, the mouth droops (causing the patient to drool saliva from the corner of his mouth), and taste perception is distorted over the affected anterior portion of the tongue. The forehead appears smooth, and the patient's ability to close his eye on the weak side is markedly impaired. When he tries to close this eye, it rolls upward (Bell's phenomenon) and shows excessive tearing. Although Bell's phenomenon occurs in normal people, it isn't apparent because the eyelids close completely and cover this eye motion. In Bell's palsy,

incomplete eye closure makes this upward motion obvious. Other symptoms may include loss of taste and ringing in the ear.

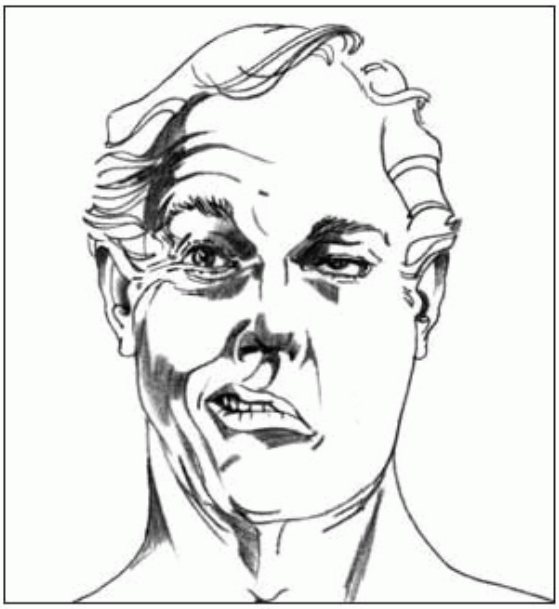
RECOGNIZING UNILATERAL BELL'S PALSY

Bell's palsy usually causes a unilateral facial paralysis. This produces a distorted appearance with an inability to wrinkle the forehead, close the eyelid, smile, show the teeth, or puff out the cheek.

DISTORTED APPEARANCE



WRINKLING THE FOREHEAD



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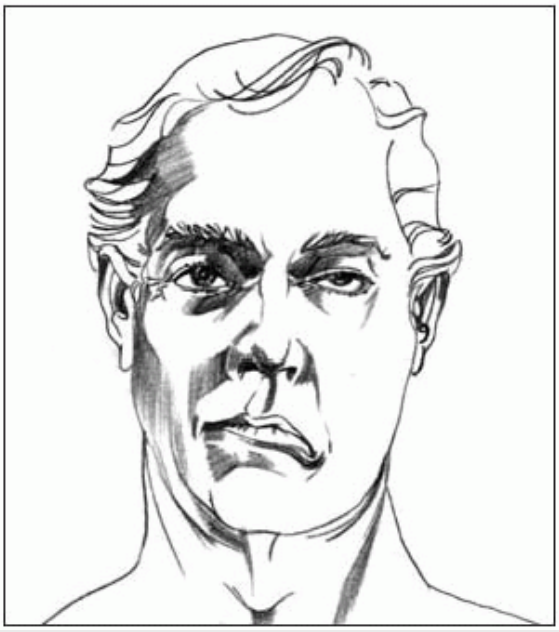

Diagnosis

Diagnosis is based on clinical presentation: distorted facial appearance and the inability to raise the eyebrow, close the eyelid, smile, show the teeth, or puff out the cheek. Electromyography helps determine

the severity of nerve damage. Blood tests may be done to rule out acute causes (sarcoidosis or Lyme disease). If no improvement is evident within several weeks of onset, magnetic resonance imaging will rule out other causes of dysfunction.

Treatment

Treatment consists of corticosteroids to reduce facial nerve edema and improve nerve conduction and blood flow. They must be given early—within 24 hours of onset of paralysis—to be most effective. Lubricants or an eye ointment may be needed to protect the eye, as well as patching during sleep.

Special considerations

Patient care includes observation for adverse drug effects, pain relief, and emotional support.

- During treatment with corticosteroids, watch for adverse effects, such as GI distress and fluid retention. If GI distress is troublesome, a concomitant antacid usually provides relief. If the patient has diabetes, frequent monitoring of serum glucose levels is necessary.
- To reduce pain, apply moist heat to the affected side of the face, taking care not to burn the skin.
- To help maintain muscle tone, massage the patient's face with a gentle upward motion two to three times daily for 5 to 10 minutes, or have him massage his face himself. When he's ready for active exercises, teach him to exercise by grimacing in front of a mirror.
- Advise the patient to protect his eye by covering it with an eye patch, especially when outdoors. Tell him to keep warm and avoid exposure to dust and wind. When exposure is unavoidable, instruct him to cover his face.
- Instruct the patient to chew on the unaffected side of his mouth. Provide a soft, nutritionally balanced diet, eliminating hot foods and fluids. Give the patient frequent mouth care, being particularly careful to remove residual food that collects between the cheeks and gums.

- Offer psychological support.

Peripheral neuritis

Peripheral neuritis (also called *multiple neuritis, peripheral neuropathy,* and *polyneuritis*) is the degeneration of peripheral nerves supplying mainly the distal muscles of the extremities. It results in muscle weakness with sensory loss and atrophy and decreased or absent deep tendon reflexes. This syndrome is associated with a noninflammatory degeneration of the axon and myelin sheaths, chiefly affecting the distal muscles of the extremities. Because onset is usually insidious, patients may compensate by overusing unaffected muscles. If the cause can be identified and eliminated, the prognosis is good.

Causes and incidence

Causes of peripheral neuritis include:

- hereditary disorders (Charcot-Marie-Tooth disease or Friedreich's ataxia)
- exposure to toxic compounds (sniffing glue or toxic compounds, nitrous oxide, industrial agents—especially solvents, and heavy metals, such as lead, arsenic, or mercury)
- infectious or inflammatory diseases (acquired immunodeficiency syndrome, botulism, Colorado tick fever, hepatitis, human immunodeficiency virus infection, leprosy, rheumatoid arthritis, sarcoidosis, systemic lupus erythematosus, diphtheria, syphilis, and Guillain-Barré syndrome)
- systemic or metabolic disorders (diabetes mellitus, dietary deficiencies [especially B_{12}], excessive use of alcohol, uremia, and cancer)
- neuropathy secondary to drugs
- miscellaneous causes (ischemia and prolonged exposure to cold temperature).

Peripheral neuropathy is common. Risk factors include diabetes, heavy alcohol use, and exposure to certain drugs and chemicals. Prolonged

pressure on a nerve (such as with a cast, splint, or other device) is also risk factor for developing nerve injury. Although it can occur at any age, incidence is highest in men between ages 30 and 50.

Complications

- Impotence
- Difficulty breathing
- Dysphagia

Signs and symptoms

The clinical effects of peripheral neuritis develop slowly, and the disease usually affects the motor and sensory nerve fibers. Symptoms vary according to which type of nerve is affected (sensory, motor, or autonomic). Neuropathy can affect any one or be a combination of all three types.

- *Sensory changes:* Damage to sensory fibers results in changes in sensation, ranging from abnormal sensations, such as burning, nerve pain, or tingling, to numbness or an inability to determine joint position in the area. Sensation changes often begin in the feet and progress toward the center of the body with involvement of other areas as the condition worsens.
- *Motor changes:* Damage to the motor fibers interferes with muscle control and can cause weakness, loss of muscle bulk, and loss of dexterity. Muscle cramping may be a sign of motor nerve involvement. Other muscle-related symptoms include lack of muscle control, difficulty or inability to move a part of the body (paralysis), muscle atrophy, muscle twitching (fasciculation) or cramping, difficulty breathing or swallowing, falling (from legs buckling or tripping over toes), or lack of dexterity (such as the inability to button a shirt).
- *Autonomic changes:* The autonomic nerves control involuntary or semivoluntary functions, such as control of internal organs and blood pressure. Damage to autonomic nerves can cause blurred vision, decreased ability to sweat (anhidrosis), dizziness that occurs when

standing up or fainting associated with a fall in blood pressure, heat intolerance with exertion (decreased ability to regulate body temperature), nausea or vomiting after meals, abdominal bloating (swelling), feeling full after eating a small amount (early satiety), diarrhea, constipation, unintentional weight loss (more than 5% of body weight), urinary incontinence, feeling of incomplete bladder emptying, difficulty beginning to urinate (urinary hesitancy), and male impotence.

Diagnosis

Patient history and physical examination delineate characteristic distribution of motor and sensory deficits. Electromyography may show a delayed action potential if this condition impairs motor nerve function. Nerve biopsy and nerve conduction tests can facilitate diagnosis.

Treatment

Effective treatment of peripheral neuritis consists of supportive measures to relieve pain, adequate bed rest, and physical therapy, vocational therapy, occupational therapy, and orthopedic interventions to promote independence, as needed. Most importantly, however, the underlying cause must be identified and corrected. For instance, it's essential to identify and remove the toxic agent, correct nutritional and vitamin deficiencies (the patient needs a high-calorie diet rich in vitamins, especially B-complex), and counsel the patient to avoid alcohol.

Over-the-counter analgesics or prescription pain medications may be needed to control nerve pain. Anticonvulsants (phenytoin, carbamazepine, and gabapentin) or tricyclic antidepressants may be used to reduce the stabbing pains that some patients experience. Whenever possible, medication use should be minimized to avoid adverse effects.

Fludrocortisone or similar medications may be beneficial in reducing postural hypotension for some patients. Medications that increase gastric motility, such as metoclopramide, are helpful for patients with reduced gastric motility.

For patients with bladder dysfunction, manual expression of urine (pressing over the bladder with the hands), intermittent catheterization, or medications such as bethanechol may be necessary.

Special considerations

Patient care includes promoting maximal independence and control of symptoms.

- Exercises and retraining may be used to increase muscle strength and control. Appliances, such as wheelchairs, braces, and splints, may improve mobility or the ability to use an affected extremity.

- The patient with decreased sensation should be taught to check his feet or other affected areas frequently for bruises, open skin areas, or other injuries. A podiatrist can usually determine whether special orthotic devices are needed.
- Discuss safety measures in the home. Safety measures for the patient experiencing difficulty with movement may include railings, specialized appliances, removal of obstacles (such as loose rugs that may slip on the floor), and other measures, as appropriate. Safety measures for the patient with diminished sensation include adequate lighting (including lights left on at night), testing water temperature before bathing or immersing the body in water, and the use of protective shoes (no open toes and no high heels). Shoes should be checked often for grit or rough spots that may cause injury to the feet.
- The patient with neuropathy (especially the patient with polyneuropathy or mononeuropathy multiplex) is prone to new nerve injury at pressure points, such as the knees and elbows. Caution him to avoid prolonged pressure on these areas from leaning on the elbows, crossing the knees, or similar positions.
- Advise the patient with orthostatic hypotension to use elastic stockings and sleep with his head elevated.
- Instruct the patient with reduced gastric motility to eat small, frequent meals and sleep with his head elevated.

- Assist the patient with bladder dysfunction with manual expression of urine and intermittent catheterization, as necessary.
- To prevent pressure ulcers, assist in turning and repositioning every 2 hours and apply a food cradle. To prevent contractures, provide range-of-motion exercises as well as arrange for the patient to obtain splints, boards, braces, or other orthopedic appliances.
- Suggest that the patient's family contact the Neuropathy Association for additional information.

PAIN DISORDERS

Complex regional pain syndrome

Complex regional pain syndrome (CRPS), also known as *reflex sympathetic dystrophy* (CRPS1) or *causalgia* (CRPS2), is a chronic pain disorder that results from abnormal healing after an injury—either minor or major—to a bone, muscle, or nerve. The development of symptoms is commonly disproportionate to the severity of the injury and seems to result from abnormal functioning of the sympathetic nervous system, the part of the nervous system that controls the diameter of blood vessels. One or more limbs and other parts of the body may be affected.

Causes and incidence

The exact cause of CRPS is unknown. Impaired communication between the damaged nerves of the sympathetic nervous system and the brain may cause interference with normal signals for sensations, temperature, and blood flow. This leads to problems in the nerves, blood vessels, skin, bones, and muscles. Infection or injury to an arm or leg may initiate CRPS. It can also occur after heart attacks and strokes. However, the condition can sometimes appear without obvious injury to the affected limb. This condition is more common in people between ages 40 and 60 but has been seen in younger people too. CRPS may also be seen in postoperative patients and in patients with diseases that can cause chronic pain, such as cancer and arthritis. Annual incidence is unknown because CRPS is often misdiagnosed. However, it has been reported in 1% to 2% of patients with various fractures and in 2% to 5% of patients with peripheral nerve injury.

Complications

- Depression
- Drug dependence

Signs and symptoms

Patients usually report severe and constant pain; severe pain is common with CRPS2

in particular. The affected area may have altered blood flow, feeling either warm or cool to the touch, with discoloration, sweating, or swelling. In time, skin, hair, and nail changes may occur along with impaired mobility and muscle wasting, especially if adequate treatment is delayed.

Diagnosis

There's no laboratory test for CRPS, so the diagnosis is based on the patient's history and clinical findings. A history of injury to an extremity may point to CRPS. Bone X-rays may aid in ruling out other conditions, such as osteomyelitis and stress fractures, which cause similar signs and symptoms. Additional tests may include bone scans, nerve conduction studies, and thermography (a test to show temperature changes and lack of blood supply in the painful area of the affected limb). With early diagnosis, prognosis improves.

Treatment

Treatment typically includes a combination of therapies such as drug therapy, with an anti-inflammatory, antidepressant, vasodilator, and analgesic used singly or in varying combinations, depending on the patient and the severity of symptoms. Steroids may be given in some patients; others may be given bone loss medications such as Actonel. Physical therapy to the injured area, application of heat and cold, the use of a transcutaneous electrical nerve stimulator unit, biofeedback, and psychological support are helpful for some patients.

Treatment may also include techniques for interrupting the hyperactivity of the sympathetic nervous system, such as nerve or regional blocks. Surgical sympathectomy—radical surgery that involves cutting the nerves to destroy the pain—may be done in severe cases; however, this method is rarely used because other sensation may be destroyed in the process.

Special considerations

- Offer emotional support to the patient and his family. Teach them about the disease.
- Monitor effects of prescribed medications.
- In addition to attending physical therapy sessions, the patient may need a home therapy regimen that includes stretching, active and passive exercises, strengthening exercises, compressive stockings or gloves to control edema, and heat or cold pack applications.
- Consult a pain care specialist to provide additional options for the patient, and help manage discomfort.
- Because chronic pain can be an emotional burden to the patient and his family, provide information on resources, such as counseling, support groups, stress-reduction methods, meditation, relaxation training, and hypnosis.

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