Chapter 3

Acute Etiologies of Neurogenic Communication Disorders

Where this icon appears, visit http://go.jblearning.com/ManascoCWS to view the corresponding video.

Whereas degenerative disorders are diseases that result from a systemic breakdown or destruction of structures within the peripheral or central nervous systems, usually from unknown or partially known mechanisms, acute etiologies include acute or traumatic events such as stroke, traumatic brain injury, seizure, tumor, surgical trauma, and infection.

Etiology The underlying cause of a symptom or deficit.

Idiopathic To be of unknown origin.

Clinicians use the term etiology to refer to the underlying cause of a symptom or deficit. Neurogenic communication disorders vary greatly in their etiologies. The most common etiologies of neurogenic communication disorders in the general population are stroke, traumatic brain injury, surgical trauma, and degenerative diseases. However, infectious disease and other conditions can also produce deficits in speech, cognition, and language. It is not uncommon for individuals with neurogenic communication disorders to have an unknown etiology of their deficits or symptoms. If an etiology is unknown or obscure, it is said to be idiopathic.

The etiologies mentioned in this chapter produce damage to the central and/or peripheral nervous system. How this damage to the nervous system manifests in deficits in communication, cognition, and behavior is determined by the site of the damage as well as the severity of the damage. Often, the site and severity of damage to the nervous system are intimately associated with the etiology. For instance, certain diseases attack specific parts of the nervous system.

It is important for speech-language pathology students to secure a basic understanding of stroke. Strokes are an overwhelmingly common condition encountered in the adult and aging populations. Before tackling the communication disorders that a stroke can produce, students must understand the

physiologic mechanisms behind this etiology and be able to recognize the early warning signs of stroke so that medical help can be acquired for the patient before permanent damage to the brain occurs.

Stroke: Cerebrovascular Accident

A stroke is the result of blood flow to a part of the brain being interrupted by a clot or a hemorrhage. Medical professionals refer to stroke as a cerebrovascular accident or, simply, CVA. Speech-language pathologists who work in hospitals and other medical settings inevitably work with many individuals who have had a stroke. Strokes can produce damage to any area of the brain or brain stem and, therefore, can create most any type of neurogenic communication disorder or cognitive deficit.

Cerebrovascular accident A stroke. An interruption of blood flow to or within the brain that permanently destroys brain tissue or causes a temporary cessation of function.

Stroke is the third leading cause of death in the United States behind heart disease and cancer (American Heart Association [AHA], 2010; Bonita, 1992). Stroke is a leading cause of hospital admission and long-term disability (AHA, 2010; Bonita, 1992). It has been estimated that every 40 seconds someone in the United States has a stroke and every 4 minutes someone dies of a stroke (AHA, 2010). Although the actual percentage of individuals experiencing communication difficulties following stroke is unknown, studies show that the presence of aphasia alone among individuals who have had strokes can be as high as 41.2% (Guyomard et al., 2009). Also, more women tend to die of stroke than men (AHA, 2010). Known factors that contribute to the likelihood of experiencing a stroke are a history of tobacco use (which doubles a person's risk for stroke), physical inactivity, atrial fibrillation, and high blood pressure (Rosamond et al., 2008).

Specifically, a stroke is when brain tissue is either permanently destroyed or temporarily ceases to function as a result of decreased or absent blood supply to the affected area. When brain tissue is permanently destroyed, the body reabsorbs the dead cells and an empty space is left on the cortex or within the brain where the tissue once was.

During a stroke, a primary source of damage to brain tissue is from the loss of oxygen supply resulting from a lack of blood supply, which transports oxygen-rich blood to the brain. The complete lack of oxygen supply to tissue is a condition known as anoxia. The partial loss of oxygen supply to tissue is hypoxia. Typically, 6 to 8 minutes without oxygen before anoxia begins to cause permanent cell death within the brain. It is therefore extremely important to be able to recognize quickly when an individual is experiencing a stroke so that appropriate medical care can be acquired.

Stroke A lesion in the brain that occurs when brain tissue is either permanently destroyed or temporarily ceases to function as a result of a lack of blood flow or a disturbance of blood supply to the affected area.

Anoxia Being completely without oxygen; an extreme lack of oxygen to brain tissue.

Hypoxia A condition in which the body lacks appropriate oxygen supply.

Ischemia Blockage of or restriction in a blood vessel.

Hemorrhage Bleeding or an open blood vessel.

There are two main forms of stroke: ischemic and hemorrhagic. The term ischemia means a blockage of or restriction in a blood vessel. The term *hemorrhagic* is derived from the word hemorrhage, which means to bleed. The majority of strokes are ischemic in nature. Typically, strokes can produce immediate deficits in cognition and language as well as weakness and difficulty seeing, hearing, and balancing.

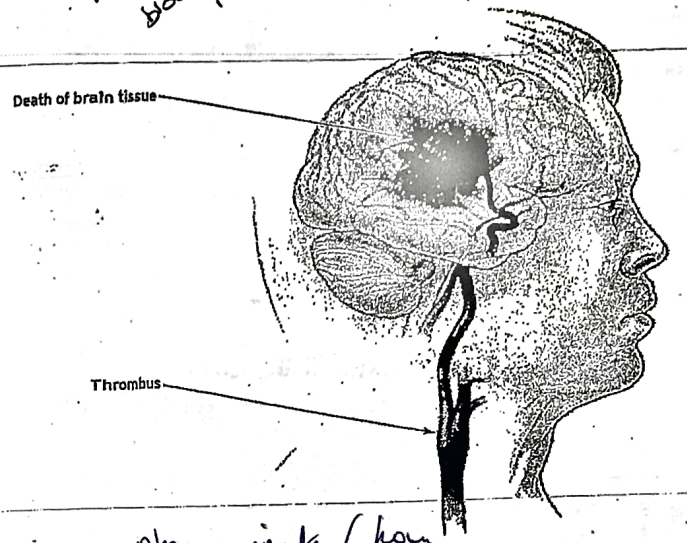


Figure 3-1 Thrombus reducing bloodflow to the brain.

Ischemic Stroke

An ischemic stroke occurs when a blood vessel supplying bloodflow to the brain becomes occluded (Figure 3-1). An occlusion in a blood vessel deprives brain tissue of the blood supply necessary for survival of the tissue. Symptoms of ischemic stroke typically develop over minutes or hours. Early warning signs or symptoms of an ischemic stroke include "loss of strength or sensation on one side of the body, problems with speech and language, or changes in vision or balance" (AHA, 2007, p. 1). See the videos *Brain Stem Stroke* and *Recovering from Stroke and Nonfluent Aphasia* for descriptions of the onset of ischemic strokes.

Ischemic stroke A cerebrovascular accident that occurs as a result of a blood vessel becoming occluded or blocked.

Thrombus A site of occlusion of a blood vessel usually the result of slow accumulation of fatty materials such as cholesterol on the walls of the artery.

There are three main forms of ischemic stroke. The first is a thrombotic stroke. A thrombus is an occlusion that forms slowly in an artery. A thrombotic stroke occurs when a thrombus interrupts blood flow to the brain, resulting in a stroke. A thrombus usually is the result of atherosclerosis. Atherosclerosis is a condition in which a person has a buildup of fatty materials such as cholesterol in the blood and this material accumulates

Thrombotic stroke A cerebrovascular accident that occurs when a thrombus forms and interrupts blood flow to the brain.

Atherosclerosis A condition in which a person has a buildup of fatty materials such as cholesterol in the blood and this material accumulates slowly on the walls of the arteries, narrowing the arteries and possibly restricting blood flow.

slowly over time on the walls of arteries, narrowing them and possibly restricting blood flow.

A mass, such as a blood clot, that originates in the body and travels through the vascular system is known as an embolus. An embolic stroke occurs when an embolus, formed elsewhere in the body, travels to the brain and lodges in a blood vessel, restricting or cutting off blood circulation within the brain. A piece of a thrombus can become an embolus (a thrombo-embolus) if, for instance, a piece of thrombus breaks off of an arterial wall and travels elsewhere within the brain to lodge and create an occlusion within a blood vessel.

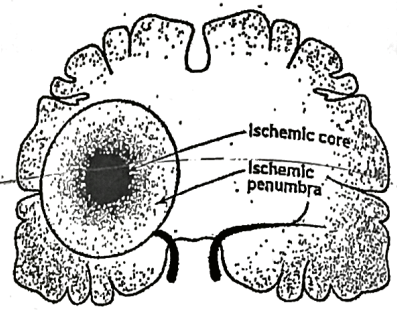


Figure 3-2 Ischemic core and penumbra.

The third type of ischemic stroke is the transient ischemic attack, known in the medical community simply as TIA and commonly as a mini stroke. A TIA is a small ischemia within the brain that resolves within 24 hours. An individual experiencing a TIA may experience mild motor and cognitive deficits that go away when the blood clot causing the ischemia is successfully broken down by the body, resolving the occlusion. Transient ischemic attacks usually do not cause permanent deficits or life-threatening health issues. However, TIAs are usually warning signs of a larger, more destructive stroke to come. Although a single TIA usually does not cause lasting deficits, multiple reoccurring TIAs can, over time, cause significant cognitive and language deficits, possibly creating what is known as vascular dementia.

Embolus A mass within the blood stream that is carried through the vascular system by the blood.

Embolic stroke When an embolus lodges within a blood vessel and restricts or cuts off blood circulation to the brain.

Thrombo-embolus When a piece of a thrombus breaks off and travels through the circulatory system, thereby becoming an embolus.

When an ischemic stroke occurs, the portion of brain tissue that is immediately deprived of the necessary level of blood flow to survive dies within an hour. This area of dead tissue is referred to as the ischemic core. The ischemic core, also known as the infarct, is the location of the focal damage within the brain following ischemia (Figure 3-2). The death of cells, or tissue necrosis, within the core is irreversible. However, circumscribing the ischemic core is the ischemic penumbra (Figure 3-2). The ischemic penumbra is the area in the brain tissue that, although it has lost the appropriate level of blood supply to function, still receives enough collateral blood flow from other vessels to stay alive. The penumbra is important because, whereas the core has experienced permanent tissue damage, the tissue within the penumbra can often be salvaged with prompt and appropriate medical treatment. This means that with timely medical intervention the brain tissue within the penumbra can be saved. Saving the penumbra improves the short-term and long-term prognoses for the patient (Sallustio, Dio-medi, Centonze, & Stanzione, 2007). Damage within the ischemic penumbra can typically be reversed within 2 to 4 hours of onset of ischemia.

Transient ischemic attack A temporary ischemia within the brain that resolves itself within 24 hours.

Ischemic core The location of the focal damage of tissue within the brain following ischemia. Also known as the infarct.

Tissue necrosis The death of body tissue.

Ischemic penumbra An area of tissue within the brain surrounding the ischemic core that receives the appropriate level of blood supply to function but that is still receiving enough collateral blood flow from other vessels to stay alive.

Hemorrhagic Stroke

A hemorrhagic stroke is when a blood vessel within the brain ruptures (hemorrhages). (See Figure 3-3.) About 13% of strokes are hemorrhagic (AHA, 2010). These strokes usually occur in individuals with high blood pressure and often during periods of high physical activity. By far the most significant risk factor associated with hemorrhagic stroke is high blood pressure, also known as hypertension. Alcohol abuse and the presence of relatives who have experienced hemorrhagic strokes also increase a person's risk. Typically, hemorrhagic strokes require immediate surgery to repair the broken blood vessel and to stop the bleeding. Individuals who experience a hemorrhagic stroke usually have less chance of survival than those who experience an ischemic stroke. However, individuals who do survive a hemorrhagic stroke often have fewer enduring deficits than those who experience ischemic strokes.

Hemorrhagic stroke A cerebrovascular accident that occurs when a blood vessel within the brain ruptures.

Hypertension High blood pressure.

Subarachnoid hemorrhage A hemorrhagic stroke that occurs when a blood vessel between the arachnoid mater and the pia mater ruptures.

There are two primary kinds of hemorrhagic stroke, the subarachnoid hemorrhage and the intracerebral hemorrhage. A subarachnoid hemorrhage is a bleed that occurs between the surface of the cerebrum and the skull. Specifically, the hemorrhage occurs in an area known as the subarachnoid space that exists between the arachnoid mater and the pia mater, which

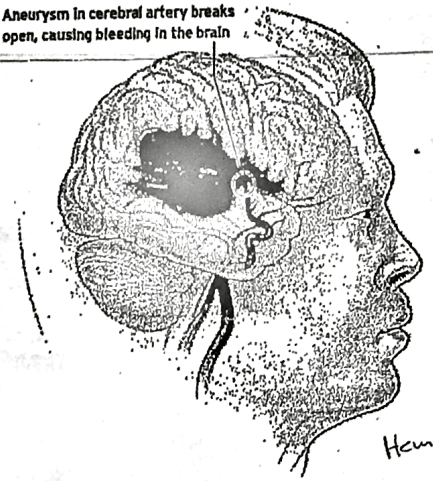


Figure 3-3 Hemorrhagic stroke.

Intracerebral hemorrhage A hemorrhage that occurs within the brain, often of traumatic origin.

Intracranial pressure The level of pressure within the skull and therefore the amount of pressure that is exerted on the brain and the amount of pressure the heart must pump against for blood to reach the brain.

overlay and protect the cerebrum. An intracerebral hemorrhage occurs when a blood vessel bursts within the brain itself.

Unlike symptoms of ischemic stroke, which can develop relatively slowly over minutes or hours, the onset of symptoms of a hemorrhagic stroke is sudden and definite. A hemorrhagic stroke is usually announced by a sudden and very severe headache, which might be associated with nausea and vomiting (AHA, 2007). Those who experience these headaches say they fall like a clap of thunder, hence their name thunderclap headaches. Although a sudden headache is considered typical of a hemorrhagic stroke, at times it can indicate an ischemic stroke.

Three primary and very dangerous mechanisms of damage to the brain are associated with hemorrhagic strokes. The first is, like ischemic stroke, when blood supply to a portion of the brain is interrupted. Unlike ischemic stroke, where blood flow simply stops because of an occlusion of a blood vessel, in hemorrhagic stroke the blood is diverted out of the burst or broken blood vessel and pours into the brain. Therefore, the second mechanism of damage following a hemorrhagic stroke is blood spilling outside of the circulatory system into the brain tissue where it does not belong, which damages the surrounding tissue that comes into contact with the blood. Finally, the continued release of blood into the brain or between the surface of the brain and the cranium increases intracranial pressure. Intracranial pressure is the level of pressure within the skull and thus the level of pressure to which the brain is exposed. Heightened levels of intracranial pressure create a very inhospitable environment for the brain. Increased intracranial pressure makes it increasingly difficult for the heart to pump blood to the brain and can quickly lead to death if not promptly treated. Many individuals have survived stroke, surgery, and worse only to die a few hours later as a result of heightened levels of intracranial pressure.

Aneurysm

An aneurysm is an abnormal stretching and ballooning of the wall of a blood vessel (Figure 3-4). An aneurysm can result from disease or hereditary factors that weaken the wall of an artery. Hypertension (high blood pressure) and atherosclerosis also can contribute by placing above-normal amounts of pressure on artery walls. Symptoms of a cerebral aneurysm are severe headache, nausea, vomiting, blurred vision or sensitivity to light, seizures, or loss of consciousness. However, there might be no symptoms at all until the aneurysm ruptures. Once a cerebral aneurysm ruptures it becomes a hemorrhagic stroke with a sudden and rapid spilling of blood into the brain. Aneurysms of the brain commonly occur within the circle of Willis (Figure 3-4). Ruptured aneurysms tend to be deadly and most of those who survive have some form of permanent disability.

Aneurysm An abnormal stretching and ballooning of the wall of a blood vessel.

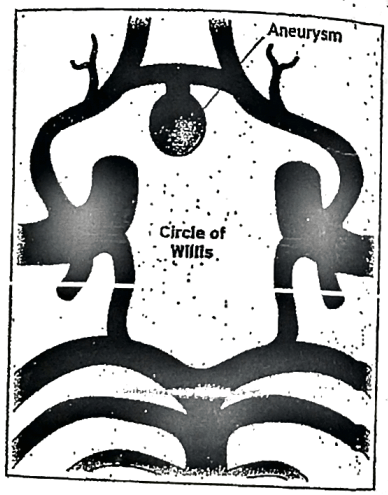


Figure 3-4 Aneurysm.

Traumatic Brain Injury

Traumatic brain injury Damage to the brain that is the result of an external and forceful event.

A traumatic brain injury (TBI) is when serious and life-threatening damage to the brain occurs as the result of an external and forceful event. This definition of TBI rules out damage to the brain as a result of disease, stroke, seizure, or surgery. Usual causes of traumatic brain injury are falls, motor vehicle and traffic accidents, being struck by an object, and violent assaults (Faul, Xu, Wald, & Coronado, 2010). The immediate impact of a TBI can range from a mild concussion to coma or death. It is estimated that about 1.4 million individuals experience a TBI each year in the United States (Faul et al., 2010).

Deficits in language and cognition following TBI are many, complex, and varied and are typically a function of which areas of the brain were damaged and to what extent. Speech-language pathologists see and treat individuals with TBI for speech, language, cognition, and swallowing disorders. The hospital-based speech-language pathologist usually begins therapy for these disorders before long-term rehabilitation efforts begin. In a best-case scenario, speech-language pathologists work with an individual with TBI from shortly after his or her admittance to the hospital all the way to the point when the individual begins returning to his or her previous life at home, work, or school. However, many of those with TBI do not make such a complete recovery and continue to live out their lives in various debilitated conditions such as coma or vegetative state.

For children with TBI, rehabilitation eventually culminates (hopefully) in the release of the child from the hospital. At this point, responsibility for continued rehabilitation of the child is passed from the in-hospital speech-language pathologist to the speech-language pathologist in the child's school. Also, because TBI is very common in children ages 4 years and younger, school-based speech-language pathologists must be familiar with TBI treatment.

Brain Tumors

A brain tumor is an abnormal growth of cells in the brain (Figure 3-5). Also known as a neoplasm, a tumor serves no purpose to the body. A primary tumor of the brain is a tumor that originates within the brain and has not spread from a tumor elsewhere in the body. A secondary tumor, or a metastatic tumor of the brain, is a cancerous tumor that spread from another part of the body to the brain. In 2009, there were an estimated 12,920 cases of brain cancer in the United States (American Cancer Society, 2009). Brain tumors are composed of the types of cellular tissue from which they originally arise. A tumor that arises within the brain is composed of abnormal growths of certain brain cells. The names given to brain tumors reflect the type of cell the tumor is composed of. For instance, cells called oligodendrocytes are myelin-producing cells in the brain

Brain tumor An abnormal growth of cells in the brain.

Neoplasm A brain tumor. An abnormal growth of cells in the brain that serves no purpose to the body.

Primary tumor A tumor that does not originate from another tumor.

Secondary tumor A cancerous tumor that spread from another part of the body. Also known as a metastatic tumor.

Metastatic tumor A cancerous tumor that spread from a primary tumor to grow in another part of the body. Also known as a secondary tumor.

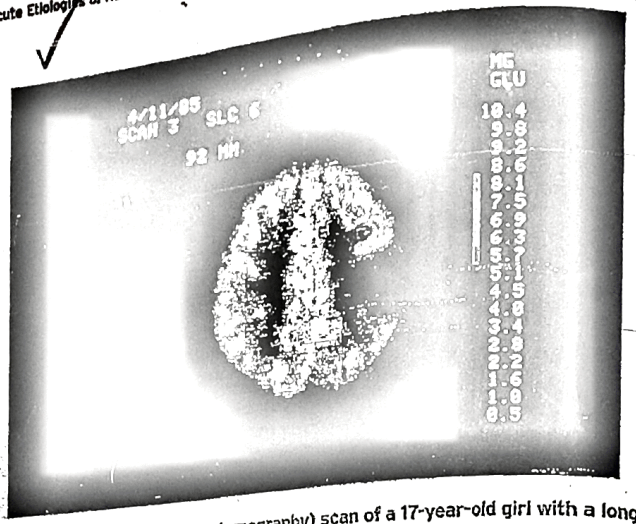


Figure 3-5 A PET (positron emission tomography) scan of a 17-year-old girl with a longstanding history of epilepsy, who has a brain tumor (the large dark spot on the right side of the image).
Source: Courtesy of Dr. Giovanni Di Chiro, Neuroimaging Center, National Institute of Neurologic/National Cancer Institute.

oligodendrocytes is termed an oligodendroglioma.

Brain tumors can be either benign or malignant. Medical doctors remove a small piece of the tumor's tissue to determine if the tumor is benign or malignant. The minor surgery to remove a piece of tissue for testing is a biopsy. A malignant brain tumor is brain cancer. Brain cancer is life threatening and can grow quickly and spread throughout the body. Brain cancer is often treated with surgical removal of the tumor followed by radiation therapy. These treatments pose their own dangers to the brain. Damage to the brain usually occurs as a result of the surgical process of cutting into the brain to remove a tumor. Radiation therapy destroys the targeted cancerous tissue in an attempt to kill a malignant tumor and cancer cells left in the area of the tumor after surgical removal. However, radiation therapy destroys healthy and cancerous cells indiscriminately; therefore, it can also damage the brain or surrounding structures. If radiation therapy is not carefully targeting the tumor, healthy tissue cells exposed to the radiation will die.

Unlike a malignant tumor, a benign brain tumor cannot spread to other parts of the body. Although the word *benign* implies a lack of possible harm, benign tumors can be quite dangerous and problematic as well. For example, even a benign tumor can grow uncontrollably within the brain, crushing an otherwise healthy brain against the skull and damaging the healthy brain tissue. When a tumor displaces or crushes areas of the brain, possibly causing

Oligodendroglioma A brain tumor arising from oligodendrocytes, which are myelin-producing cells within the brain.

Biopsy Surgery to remove a piece of tissue for diagnostic purposes.

Malignant brain tumor Brain cancer.

Brain cancer A brain or one located centrally within the spinal cord.

Benign brain tumor A tumor within the brain that cannot spread to other parts of the body.

Mass effect The displacement effect or crushing force on nearby tissues that a tumor exerts.

Surgical trauma The collateral damage to the tissues of the body that occurs during the process of surgery.

symptoms, it is said to be creating a mass effect. As mentioned previously, surgical intervention to remove a benign tumor can itself be damaging to the brain and can produce even more complications and deficits than the tumor itself, though the individual's life is preserved.

Brain tumors can produce focal damage within the brain as a stroke might, but usually present with different symptoms. Deficits produced by a tumor depend on the area of the brain the tumor affects and to what degree. However, headache is a common symptom. Whereas symptoms of stroke occur somewhat suddenly, symptoms of tumor occur and worsen gradually over longer periods of time as the tumor grows.

Surgical Trauma

Surgery is performed on a person's brain for various reasons. Surgery on the brain is often performed to remove a tumor or repair a hemorrhagic stroke. Surgery on the brain is never lightly considered and is usually undertaken only in very serious or life-threatening circumstances. Brain surgery often results in surgical trauma. Surgical trauma is the collateral damage to the delicate tissues of the brain that occurs during the process of achieving the objective of the surgery. Surgical trauma is often a necessary consequence of removing a tumor or repairing a bleed to save the patient's life. Unfortunately, once these individuals are medically stable, a speech-language pathologist might need to evaluate and treat them for acquired language, cognitive, speech, or swallowing deficits. Medical professionals must also try to manage the secondary risks of brain surgery: seizures, additional cerebrovascular accidents, acquired infections, and increased intracranial pressure.

See the *Surgical Trauma to the Vagus* video for a discussion of related material.

Infection

Many infections are capable of damaging the central nervous system (CNS) and the peripheral nervous system (PNS). Infections can be viral, fungal, bacterial, or parasitic. The impact an infection has on cognition, language, or motor movement (i.e., speech and swallowing) depends on the site of the infection, the nature of the infection, and the extent of damage created by the infection. Some infections might affect only the CNS and create alterations and deficits in cognition and language. Others affect only the PNS, affecting motor control of the body, which affects speech. Some infections affect both the CNS and PNS. The diseases addressed in the following subsections, selected for their clinical as well as historical and educational significance, are but a few that involve the human nervous system.

Encephalitis

Encephalitis is a general term indicating an acute infection and/or inflammation of the brain or spinal cord. There are many different forms of encephalitis. Generally, encephalitis is caused by a viral, bacterial, or parasitic infection of the brain or spinal cord. The symptoms often reflect the type as well as location of the infection. Symptoms often include headaches, fevers, confusion, and seizures.

Encephalitis A general term that refers to an acute infection and/or inflammation of the brain or spinal cord.

A particularly interesting form of encephalitis that made its way into public awareness is encephalitis lethargica. In the early part of the 20th century, there was an epidemic of this disease. This outbreak of encephalitis lethargica occurred alongside and was possibly the result of a simultaneous influenza epidemic

(Mortimer, 2009). Known as the sleepy sickness in the 1910s and 1920s, first signs of the disease are a fever accompanied by a low level of arousal or sleep state. Eventually, this disease leads to muscle rigidity and weakness, disturbances in initiating movement, and cardiac and respiratory abnormalities. Encephalitis lethargica is fatal in approximately one-third of its victims. It causes inflammation and damage to subcortical structures that are responsible for autonomic processes such as the sleep/wake cycle, heart rate, breathing rate, and the control of movement. Specifically, these structures are the midbrain, basal ganglia, and substantia nigra (Anderson, Vilensky, & Duvoisin, 2009). Survivors of this disease usually display distinctly Parkinsonian features as a result of the damage to their subcortical structures. These symptoms can include difficulty initiating and controlling volitional movement as well as inhibiting nonvolitional movement.

Encephalitis lethargica was virtually forgotten by modern medicine until the publication of Awakenings by Oliver Sacks in which he details his administration of the then-experimental drug levadopa (L-Dopa) during the 1960s to survivors of the 1920s encephalitis lethargica epidemic. Sacks (1990) wrote of the survivors of the 1920s epidemic who were still living and had been placed, years earlier, in a long-term care hospital where he worked. He described these patients as being alive, yet not fully awake, and unable to move or speak. The patients displayed an extreme form of Parkinsonism resulting from the damage the disease wrought in their subcortices. Although no large-scale outbreaks of encephalitis lethargica have occurred since the 1920s, sporadic cases are still regularly diagnosed and reported (Anderson et al., 2009).

A rare and idiopathic form of encephalitis is Rasmussen's encephalitis. Rasmussen's is a fast-moving encephalitis characterized by T cells of the immune system attacking and causing inflammation in either the right or left cerebral hemisphere. As a result of this inflammation, the presenting symptom of Rasmussen's encephalitis is seizure activity that creates a unilateral tremor in an extremity contralateral to the affected cerebral hemisphere. Individuals with Rasmussen's encephalitis progress over the course of months into seizures that grow more severe as more of the diseased cerebral hemisphere is affected. Treatment is medical to control seizure activity, but seizures ultimately grow so debilitating and life threatening that individuals with this disease must undergo surgery to remove large portions of the affected cerebral hemisphere or even the entire cerebral hemisphere. This surgery is known as a hemispherectomy. Hemispherectomy is most often performed to remove large tumors existing in a single cerebral hemisphere or to control intractable seizure activity, as in conditions such as Rasmussen's encephalitis. See the video *Rasmussen's Encephalitis, Seizures, and Hemispherectomy* for an interview with an individual who underwent a hemispherectomy for the remediation of Rasmussen's encephalitis.

HIV/AIDS

Human immunodeficiency virus (HIV) is the virus that leads to AIDS (acquired immune deficiency syndrome). HIV/AIDS can be transmitted through sexual contact, through blood, or from an infected mother to her child. HIV slowly weakens the immune system until another pathogen overtakes the individual's body, leading to death. HIV/AIDS is a pandemic, and no known cure for the disease exists. In 2007, the World Health Organization (WHO) estimated that there were 32.2 million people infected with HIV (UNAIDS, 2007). That same year, the World Health Organization estimated that 2.1 million people died of AIDS (Joint United Nations Programme on HIV/AIDS [UNAIDS] & World Health Organization [WHO], 2007), including 330,000 children.

Human immunodeficiency virus (HIV) The virus that leads to AIDS (acquired immune deficiency syndrome).

Acquired immune deficiency syndrome (AIDS) The final stage of HIV disease characterized by severe damage and impairment of the immune system.

neuroAIDS Neurological changes as a result of HIV/AIDS that create cognitive deficits and dementia. Also known as HIV/AIDS dementia and HIV-associated neurocognitive disorder (HAND).

AIDS is known to cause neurologic changes and deficits. Neurologic symptoms can occur before an individual knows he or she is infected (Singer, Valdes-Sueiras, Commins, & Levine, 2010). However, until recently, individuals with AIDS often did not live long enough for these cognitive and motor deficits to be a concern. With the advent of more effective medical treatments in the form of combinations of medicines known as drug cocktails, individuals with HIV are living long enough to experience these neurologic symptoms, often referred to as neuroAIDS or HIV/AIDS dementia (Gendelman, Grant, Everall, Lipton, & Swindells, 2005) and more recently as HIV-associated neurocognitive disorder (HAND) (Singer et al., 2010). The term HIV/AIDS dementia is applied only if neurocognitive deficits are severe enough to affect the individual's daily life. The most common neurocognitive changes seen in AIDS include impairments in ability to learn new information, loss of gross and fine motor abilities including gait disturbances, reduced attention abilities, slowness in processing information, disfluent speech, and impaired recall (Gendelman et al., 2005; Singer et al., 2010). Although language is often unaffected, mild language deficits and severe deficits in functional use of language in individuals with HIV/AIDS dementia have also been reported (McCabe, Sheard, & Code, 2008).

HIV/AIDS dementia Cognitive changes as a result of HIV/AIDS that are severe enough to affect an individual's activities of daily living.

Creutzfeldt-Jakob Disease

In the 1920s, Hans Creutzfeldt and Alfons Jakob documented a degenerative and fatal brain disease. This illness came to be known as Creutzfeldt-Jakob disease and has genetic as well as infectious etiologies (Praveen, Sinha, Chandrasekhar, Vijayan, & Taly, 2006). It is currently believed that most cases are produced by a type of infectious pathogen known as a prion. A prion is a small infectious protein with its own genetic coding. Although debatable, it is commonly believed that the prion disease known in animals as mad cow disease produces a variant of Creutzfeldt-Jakob disease in humans.

Prion A small infectious protein with its own genetic coding that attacks structures within the central or peripheral nervous system.

Myoclonus An involuntary rapid twitching of a muscle or group of muscles.

Creutzfeldt-Jakob disease attacks the central nervous system, currently has no known cure, and is usually fatal within a year of onset of symptoms. General symptoms of this disease include dementia with rapid onset and involuntary movement disturbances called myoclonus. Myoclonus is the involuntary rapid twitching of a muscle or group of muscles.

Praveen and colleagues (2006) document the dementia symptoms of a 69-year-old woman with Creutzfeldt-Jakob disease as including behavioral abnormalities, emotional volatility in the form of inappropriate anger or crying, and "irrelevant talk" (p. 418). One month later, this woman developed slurred speech, gait abnormalities, and "excessive sleepiness" (p. 418). She later developed myoclonus in her limbs and became bedbound.

Creutzfeldt-Jakob disease has also been noted to cause akinetic mutism (Gozke, Erdal, & Unal, 2008), which is an inability to speak as a result of being unable to move. Saigusa and colleagues (2008) document a case of bilateral paralysis of the vocal folds in a case of Creutzfeldt-Jakob disease. Certain Alzheimer-like neuropathologic changes are present in the brain tissue of those affected by Creutzfeldt-Jakob disease. One of these is the occurrence of amyloid plaques (Figure 3-6).

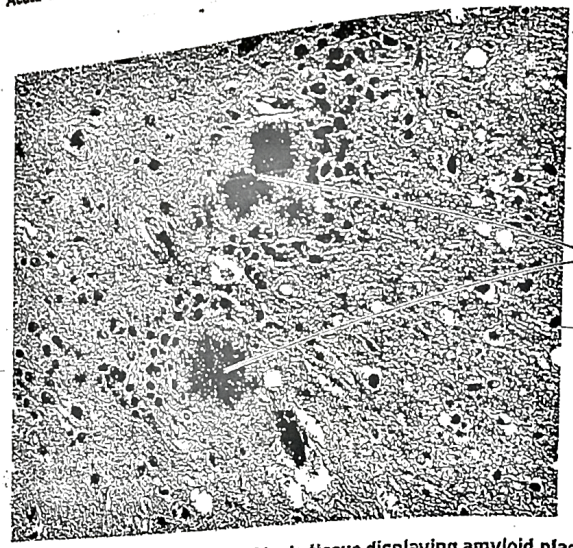


Figure 3-6 Stained photomicrograph of brain tissue displaying amyloid plaques created by variant Creutzfeldt-Jakob disease.

Source: Courtesy of Teresa Hammett, Department of Health and Human Services Centers for Disease Control and Prevention.

Syphilis

Syphilis is a sexually transmitted disease that is highly treatable with the antibiotic penicillin and is curable. This disease is caused by corkscrew-shaped bacteria called spirochetes. The spirochete that causes syphilis is *Treponema pallidum* (Figure 3-7). Syphilis infection initially causes an open sore at the site of initial infection. Symptoms then progress to include ulcers, rashes, fevers, and headaches. If left untreated, symptoms become more severe, progressing to anemia, liver and heart disease as well as disease of the nervous system.

Syphilis A sexually transmitted disease that is caused by corkscrew-shaped bacterium called spirochetes. Syphilis is highly treatable and curable with the antibiotic penicillin.

Neurosyphilis A variation of syphilis that infects the nervous system.

A specific variation of syphilis that affects the nervous system is neurosyphilis. Neurosyphilis usually occurs years after an individual is initially infected and treated for a primary infection of syphilis. However, infiltration of the CNS by syphilis can begin as early as a few days or weeks following infection (Ho & Lukehart, 2011). Neurosyphilis can present with an array of neurologic signs and symptoms including meningitis, headache, stiff neck, changes in vision or visual abnormalities, facial weakness, cognitive deficits, motor problems, and involvement of individual cranial nerves (Ho & Lukehart, 2011; Jacob et al., 2005). Intensive penicillin therapy is used to treat neurosyphilis. The earlier the treatment is provided, the better the prognosis for recovery.

Simultaneous infection of both syphilis and HIV is high among individuals with syphilis (Ho & Lukehart, 2011). This is because syphilis causes open sores on or around the genitalia. HIV, also a sexually transmitted disease, can then be transmitted more easily through these open sores. Furthermore, an immune system suppressed by HIV can make infection with syphilis a far more dangerous and serious condition than usual.

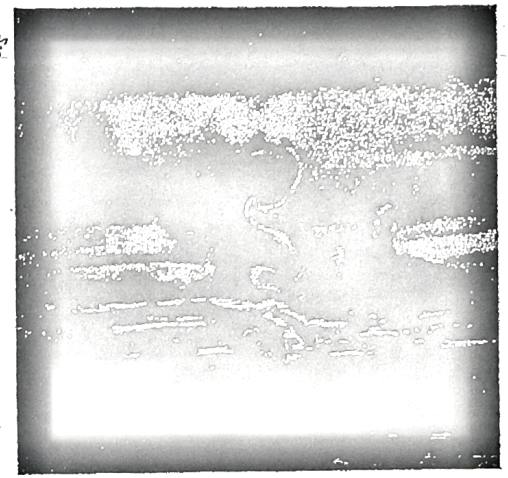


Figure 3-7 Micrograph of *Treponema pallidum*.

Source: Courtesy of David Cox, Department of Health and Human Services Centers for Disease Control and Prevention.

Poliomyelitis

Poliomyelitis, commonly known simply as polio, is caused by a virus. Polio is highly preventable with a vaccine. Polio reached epidemic proportions in the United States during the early 20th century prior to development of an effective vaccine (Cono & Alexander, 2002). The polio virus primarily attacks children and is transmitted primarily by fecal matter in drinking water. Although polio is now rare (though still present) in the United States and United Kingdom, it is still an active threat in many developing countries. The polio virus attacks efferent nerve tracks of the PNS. For an interview with a woman who experienced polio as a child see the video *Poliomyelitis*. Polio is usually categorized by the sections of the PNS that it affects. Polio affecting the spinal nerves and the muscles innervated by the spinal nerves is referred to as spinal polio. Polio affecting the cranial nerves and the muscles innervated by the cranial nerves is referred to as bulbar polio. If both spinal and cranial nerves are affected, it is termed bulbospinal polio and affects all muscles of the body to some degree.

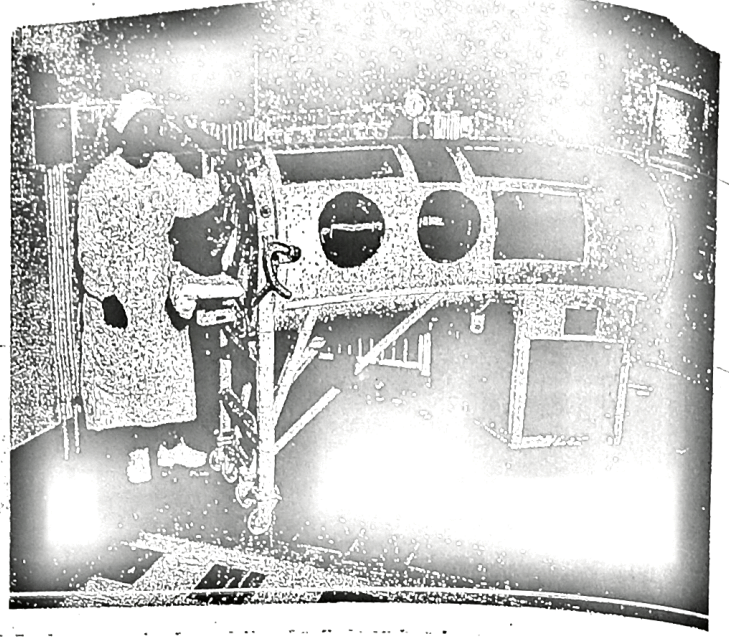
Characteristic symptoms of polio are nonsymmetrical paralysis with diminished or absent reflexes (Cono & Alexander, 2002). The paralysis and weak reflexes reflect the inability of the PNS to deliver volitional and nonvolitional commands of movement from the CNS to the muscles. The machine known as

Poliomyelitis A virus that attacks the PNS and causes paralysis and absent reflexes. Also known as polio.

Spinal polio Poliomyelitis that affects the spinal nerves and the muscles innervated by the spinal nerves.

Bulbar polio Poliomyelitis that affects the cranial nerves and the muscles innervated by the cranial nerves.

Bulbospinal polio Poliomyelitis that affects both spinal and cranial nerves and the muscles they innervate.



Source: Courtesy of Department of Health and Human Services Centers for Disease Control and Prevention.

Post polio syndrome Symptoms of muscle weakness, muscle pain, and fatigue within those limbs that were affected by a previous polio infection.

the Iron Lung (Figure 3-8) was once a common sight in hospitals and was responsible for enabling patients with severe polio to breathe despite the paralysis of their respiratory muscles. Polio is fatal in 2–10% of cases. Major recovery of muscle function usually occurs within 6 months of infection, but symptoms can persist indefinitely and even reemerge in later life as post polio syndrome (Cono & Alexander, 2002).

Post polio syndrome arises in about 25–50% of individuals who recover from a previous infection with polio. It is characterized by muscle weakness, muscle pain, and fatigue of the limbs that were affected by the initial polio infection. Post polio syndrome can affect facial muscles involved in speech as well as muscles of the neck and larynx, resulting in difficulties with voice production and swallowing (Silbergleit, Waring, Sullivan, & Maynard, 1991; Soderholm, Lehtinen, Valtonen, & Ylinen, 2010). See the video *Post Polio Syndrome* for an interview with an individual living with this disorder.

Seizures

Certain infectious diseases and congenital disorders (such as epilepsy, of which there are many forms) produce seizures. However, seizures also occur as a result of stroke, traumatic brain injury, tumor, or surgical trauma to the brain, and as such it is important for speech-language pathologists to understand and recognize them.

Seizure A sudden, often periodic, abnormal level of electrical discharge occurring within the brain.

Aura The period of time immediately preceding the full onset of a seizure, in which a person might experience some warning signs that a seizure is imminent.

Ictus The main stage of the seizure during which the primary symptoms are experienced.

Post ictus The stage of seizure that follows the ictus and that can last for minutes or hours and during which people might display lethargy and confusion and experience memory loss, weakness, and depression.

Post-ictal confusion The short-lived cognitive deficits following the ictus stage of a seizure.

Interictal period The time between the end of one seizure and the beginning of the next seizure.

Status epilepticus A state of constant seizure. When an individual experiences one seizure that leads right into another seizure with no interictal period.

The brain uses electricity to communicate with itself and the rest of the nervous system. Neurons generate and send electrical signals to each other during normal operation. Electrical impulses constantly fly among the billions of neuronal connections in the brain. However, if too much electricity occurs in the brain, a seizure will result. A seizure is a sudden, often periodic, abnormal level of electrical discharge in the brain. Seizures are most succinctly described as storms of electricity in the brain.

Seizures can be mild in nature and produce only a slowly accumulating level of brain damage in the affected areas over time. Or they can be far more severe with the potential of a single seizure producing immediate and permanent brain damage and even death.

There are many different categorizations of the stages of seizure. By most accounts, the three primary stages include the aura, ictus, and post-ictus. These stages are most evident in severe seizures. The aura is the period of time immediately preceding the full onset of a seizure in which a person might experience some warning signs that a seizure is imminent. Signs of oncoming seizure are varied and include headache, déjà vu, panic, nausea, radical mood shifts, tingling in the limbs, or visual abnormalities. The ictus is the main stage of the seizure when the person experiences the primary symptoms. In a tonic clonic seizure (discussed later), this is the period when the individual loses consciousness and begins convulsing until the motor activity ceases and the affected individual begins to regain consciousness. After regaining consciousness, the individual enters the post-ictus stage. The post ictus is the period that comes after the ictus. It can last for minutes or hours. During the post ictus, individuals are often lethargic and confused, and they might experience memory loss, weakness, and depression. This stage is often characterized by post-ictal confusion, which is a short-term cognitive deficit. The time between seizures (after the post ictus and prior to the next aura) is known as the interictal period. Status epilepticus is when a person experiences seizures, one after another that lead directly into each other with no interictal period. Status epilepticus is a severe and life-threatening condition that very negatively affects quality of life.

This text contains two descriptions of cases involving status epilepticus. In both cases, radical surgery was performed to reduce seizure activity. The first is the clinical note of patient H.M., who had his hippocampi removed to reduce seizures but was left with profound memory deficits. The second is of a woman with Rasmussen's encephalitis who had her right cerebral hemisphere removed to stop the hundreds of seizures she was having each day. Go to the video *Rasmussen's Disease, Seizures, and Hemispherectomy* to hear this woman describe her problem, her seizures, and the surgery that saved her life and allowed her to grow up and become (of all things) a speech-language pathologist.

The kind of seizure experienced depends on which part of the brain is affected. Specialists typically categorize seizures as partial or generalized.

Partial seizure A seizure in which pathologic levels of electricity in the brain remain confined to a particular region of the brain.

Simple partial seizure A partial seizure during which the seizure activity within the brain is limited to a small area within one cerebral hemisphere and the individual experiencing the seizure remains conscious.

Complex partial seizure A seizure that occurs over a large section of a single cerebral hemisphere and that creates an altered state of consciousness.

Generalized seizures A seizure that affects the entire brain and is associated with a total loss of consciousness.

Tonic clonic seizure A seizure in which an individual passes through a stage of muscle contraction and loss of consciousness (tonic phase) followed by a phase of abnormal motor activity (clonic phase).

Tonic phase The initial phase of a tonic clonic seizure characterized by a sudden stiffening of the body and limbs due to muscle contractions and a loss of consciousness.

Partial Seizures

Individuals with epilepsy most commonly experience partial seizures. During a partial seizure, the pathologic levels of electrical activity are confined to a particular region of the brain. Partial seizures can be considered pathologic overstimulation of a certain part of the brain created by an excess of electrical activity in the affected area. Partial seizures can create just about any imaginable motor, sensory, or emotional symptom because these seizures can occur in any part of the brain, though some areas are far more prone to seizure than others.

The symptoms an individual experiences during a partial seizure depend on which area of the brain is seizing. For example, a partial seizure in the occipital-parietal visual processing areas of the brain can produce visual hallucinations or somatosensory and visual abnormalities. Similarly, a seizure isolated to the temporal lobes might produce auditory hallucinations, such as the sound of music, if they occur in the area of the temporal lobes dedicated to processing music. Sacks (1970) describes a case of partial seizures of the temporal lobe in which an elderly woman wakes in the middle of the night to find herself experiencing the auditory hallucination of a deafening string of Irish songs from her youth playing over and over. However, some patients hear environmental sounds such as doors slamming or glass breaking.

There are two primary forms of partial seizure, simple partial seizures and complex partial seizures. In a simple partial seizure, the seizure activity in the brain is limited to a small area in one cerebral hemisphere and the individual experiencing the seizure remains conscious. If a seizure occurs over a larger section of a single cerebral hemisphere and creates an altered state of consciousness, the seizure is categorized as a complex partial seizure. Complex partial seizures often create changes in level of awareness or result in a dreamlike state. Individuals might produce some speech or mumbling and perhaps execute some seemingly purposeful movement. Nonetheless, during a complex partial seizure speech makes little sense and movement is highly disorganized. Complex partial seizures might also lead to a generalized seizure.

Generalized Seizures

Whereas partial seizures affect a localized portion of the brain, generalized seizures affect the entire brain and are associated with total loss of consciousness or awareness. There are two forms of generalized seizure: the tonic clonic seizure (once known as the grand mal) and the petit mal seizure, also known as absence attacks.

The tonic clonic seizure is so named because it manifests two distinct phases during the ictus. These two phases are the tonic phase and the clonic phase. The tonic phase at the beginning of the ictus signals the onset of the seizure, and the affected individual loses consciousness and the body stiffens as a result of muscle contractions. The limbs might be rigidly stiffened toward the body or extend away. If the individual is standing, he or she will fall. The mere risk of fall associated with tonic clonic seizures can be a serious threat. Often, the onset of the tonic clonic seizure is announced by an automatic loud moan or even a ragged scream as the vocal folds are adducted and air is simultaneously forced between the adducted vocal folds by the contracting thoracic muscles. The tonic phase is short, lasting only a few seconds, and leads into the clonic phase.

Clonic phase The second phase of a tonic clonic seizure characterized by a shaking, jerking, extraneous body movement.

Petit mal seizure A generalized seizure in which an individual loses awareness for a few seconds and might seem simply to stare off into space before coming to.

During the clonic phase, the individual begins to convulse and displays often violent twitching of the extremities and shaking of the entire body. The clonic phase usually lasts 2 to 3 minutes during which motor activity slowly decreases and leads to the post ictus. Tonic clonic seizures are almost always followed by a period of post-ictal sleep. Post-ictal confusion and amnesia are present when the individual regains consciousness and slowly wear off.

Whereas the chances of not recognizing the presence of a tonic clonic seizure are remote, petit mal seizures often go undiagnosed for years. The petit mal seizure, or absence attack, is a generalized seizure in which an individual loses awareness for a few seconds and seems simply to stare into space before coming to. There is no gross motor activity, shaking, or convulsing as seen in the tonic clonic seizure. In fact, parents and school teachers often mistakenly assume children who experience petit mal seizures are simply daydreamers and attribute their poor academic performance to not paying appropriate attention. In reality, the child is seizing and experiencing only bits and pieces of what is occurring in the classroom. During the time that these seizures go undiagnosed, they are also not being managed medically and are totally uncontrolled. Over time, these uncontrolled seizures can cause significant damage to the child's brain and lead to cognitive, language, and motor deficits.

Author's Note

How to Help a Person Experiencing a Seizure

If you are with someone who has lost consciousness as a result of a seizure, you should know a few simple items that will help you keep the person safe. First, be sure you never put anything in the mouth of an individual experiencing a seizure. Do not try to restrict the person's movements. Also, be sure to clear away any dangerous or sharp objects around the individual experiencing a seizure. Turn the person onto his or her side to keep saliva from falling into the airway, and place something soft under the individual's head, such as a pillow or a shirt. Finally, stay with the person until the seizure ends.

If you are a nonmedical professional, such as a speech-language pathologist, working in a medical setting and you are with a person who begins seizing, usually the most appropriate action is to get medical help immediately. Most often this takes the form of calling loudly for a nurse or doctor while staying with the patient. Protocols vary among institutions.

Main Points

- An etiology is the underlying medical cause of a symptom or deficit. An idiopathic etiology is one of unknown origin.
- Neurogenic communication disorders usually result from damage to the CNS and/or the PNS. Such damage can cause communicative, cognitive, language, and/or behavioral deficits. Etiologies of neurogenic communication disorders are often stroke, traumatic brain injury (TBI), surgical trauma, degenerative disorders, and infectious diseases.
- A stroke occurs when blood flow to a part of the brain is interrupted by a clot or hemorrhage within the brain. A stroke is also known as a cerebrovascular accident (CVA). There are two main categories of stroke: ischemic and hemorrhagic.
- Ischemic strokes occur when a blood vessel supplying blood to the brain is blocked. This blockage deprives the brain tissue of blood supply necessary to survive. There are three main forms of ischemic stroke: thrombotic, embolic, and transient ischemic attacks.
  - A thrombotic stroke is when an occlusion forms within a blood vessel and restricts blood flow to the brain. An occlusion that forms in a cumulative fashion and restricts blood flow to brain tissue is known as a thrombus. A thrombus is usually a result of atherosclerosis.
  - An embolic stroke is when a mass traveling through the vascular system (an embolus) lodges in a blood vessel usually inside the brain and restricts blood flow to brain tissue. A thrombus can become an embolus if any piece of it breaks off the wall of an artery and travels to lodge elsewhere and create a blockage of blood flow to the brain.
  - Transient ischemic attacks (TIAs), also known as mini strokes, are when a small ischemia within the brain occurs and is resolved by the body within 24 hours. TIAs do not cause permanent damage; however, recurring TIAs can cause language and cognitive deficits. TIAs can also be a warning sign of a larger oncoming stroke.
- Hemorrhagic strokes occur when a blood vessel within the brain ruptures.
- Three mechanisms of damage to the brain are possible with hemorrhagic strokes. First, the blood supply to a portion of the brain is interrupted as a result of the broken or burst blood vessel. Second, the blood from the hemorrhaged vessel spills outside the circulatory system into the brain and damages the tissue it comes into contact with. Third, intracranial pressure increases because of the continued release of blood into the brain or between the skull and the cranium.
  - There are two types of hemorrhagic strokes: subarachnoid and intracerebral.
  - Subarachnoid hemorrhagic strokes occur when there is a bleed between the surface of the brain and the skull in an area known as the subarachnoid space.
  - Intracerebral hemorrhagic strokes occur when a blood vessel bursts within the brain itself.
- An aneurysm is the abnormal stretching or ballooning of an arterial wall. Aneurysms can be the result of hypertension, disease, hereditary factors, or atherosclerosis. When an aneurysm ruptures, it becomes a hemorrhagic stroke. Aneurysms often occur within the circle of Willis.
- A traumatic brain injury (TBI) is when serious and life-threatening damage to the brain occurs as a result of an external and forceful event. A TBI is usually the result of a forceful event such as a fall, motor vehicle accident, violent assault, sports-related accident, or being struck on the head by an object. The language and cognitive deficits resulting from a TBI are complex and vary depending on which areas of the brain are damaged and to what extent.
- A brain tumor is an abnormal growth of cells in the brain. A brain tumor is also known as a neoplasm. A primary tumor of the brain is a tumor that originates in the brain. A secondary tumor of the brain is a cancerous tumor that spreads from another part of the body to the brain. A secondary tumor is also called a metastatic brain tumor. The deficits produced by a brain tumor depend on the area of the brain the tumor affects and to what degree. A malignant brain tumor is brain cancer.
  - Surgical trauma is damage to the delicate tissues of the brain that might occur with the surgical removal of a tumor or the repair of a hemorrhage in the brain to save a person's life. Those who have surgical trauma can experience acquired language and cognitive deficits.
  - Infections can also cause damage to the CNS and PNS. Infections can be viral, fungal, bacterial, or parasitic. The deficits caused by infections depend on the site of the infection, the nature of the infection, and the extent of damage done by the infection. A multitude of infections of the nervous system can affect speech, language, and/or cognition. Some of these infections include encephalitis, HIV/AIDS, Creutzfeldt-Jakob disease, syphilis, and poliomyelitis.
    - Encephalitis is a general term for an acute inflammatory infection of the brain or spinal cord. A viral or bacterial infection of the brain or spinal cord causes encephalitis. The symptoms of encephalitis vary depending on the type and location of the infection.
    - Human immunodeficiency virus (HIV) leads to acquired immune deficiency syndrome (AIDS). HIV/AIDS can cause neurologic changes and deficits, which are known as neuroAIDS, HIV/AIDS dementia, or HIV-associated neurocognitive disorder. Some deficits include inability to learn new information, slow information processing, disfluent speech, impaired recall, and reduced attention ability. Mild to severe deficits in the use of functional language might also occur.
    - A small infectious protein called a prion causes Creutzfeldt-Jakob disease. Prion diseases, including Creutzfeldt-Jakob disease, attack the CNS. The symptoms include dementia with rapid onset and involuntary movement disturbances called myoclonus. Certain Alzheimer-like neuropathologic changes, such as amyloid plaques, are present in the brain tissue of those affected by Creutzfeldt-Jakob disease.
    - Syphilis is a sexually transmitted disease caused by a corkscrew-shaped bacterium called a spirochete. Neurosyphilis is a variation of syphilis that affects the nervous system. Some signs and symptoms of neurosyphilis include meningitis, visual difficulties or abnormalities, facial weakness, cognitive deficits, and motor problems.
  - Poliomyelitis is caused by a virus that attacks the motor nerve tracks of the PNS. Symptoms of polio include nonsymmetrical paralysis with diminished or absent reflexes.
  - Seizures are sudden, often periodic, and abnormal levels of electrical discharge in the brain.
    - The three primary stages of a seizure are the aura, ictus, and post ictus. The aura is the period immediately before the seizure during which a person might experience warning signs of an upcoming seizure. The ictus is the main stage of the seizure that can include loss of consciousness and convulsions. The post ictus is the period right after the ictus during which a person can experience confusion, memory loss, weakness, and/or depression. The time between seizures is called the interictal period. Status epilepticus is when a person experiences seizures without an interictal period. There are main two categories of seizures: partial seizures and generalized seizures.

- Partial seizures occur when the abnormal levels of electrical activity remain within a particular region of the brain.
- The two primary forms of partial seizures include simple partial seizures and complex partial seizures. During a simple partial seizure, the affected individual remains conscious and the seizure is restricted to a limited region of the brain. During a complex partial seizure, the individual seizing experiences altered states of consciousness and larger or multiple regions of seizure activity occur in one single cerebral hemisphere.
- Generalized seizures occur when the abnormal levels of electrical activity affect the entire brain. A total loss of consciousness or awareness occurs with generalized seizures.
- There are two forms of generalized seizures: tonic clonic seizures and petit mal seizures. During tonic clonic seizures, the individual loses consciousness and the body stiffens and convulses. During petit mal seizures, an individual loses awareness for a few seconds and might assume the posture of a daydreamer or an absent stare. Petit mal seizures are usually a disorder of childhood.

Review Questions

1. Why are some etiologies called idiopathic?
2. Compare and contrast how ischemic and hem-
3. What are the three main forms of ischemic strokes and how do they differ?
4. Why is saving the ischemic penumbra a priority for medical professionals?
5. What are the two main forms of hemorrhagic stroke and how do they differ?
6. What is an aneurysm and why is having one dangerous?
7. What is a traumatic brain injury? What are some common causes of traumatic brain injury?
8. How do primary tumors and secondary tumors differ?
9. How might a benign tumor cause damage to the brain?
10. What is encephalitis? Give one example of encephalitis and what its effect on speech, language, or cognition might be.
11. How might HIV/AIDS affect speech, language, or cognition?
13. What is syphilis, what organism causes this disease, and what are some ways it can affect speech, language, or cognition?
14. What is a seizure?
15. Name and describe the three primary stages of seizure.
16. What is the term that describes a state of constant seizure?
17. How are simple partial seizures and complex partial seizures different?
18. How are generalized seizures different from partial seizures?
19. How are tonic clonic seizures different from petit mal seizures?
20. Describe how you would assist/help a person experiencing a tonic clonic seizure.

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