

# THE MERCK MANUAL HOME EDITION



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## Encephalitis

Encephalitis is inflammation of the brain that occurs when a virus directly infects the brain or when a virus, vaccine, or something else triggers inflammation. The spinal cord may also be involved, resulting in a disorder called encephalomyelitis.

- People may have a fever, headache, or seizures, and they may feel sleepy, numb, or confused.
- Magnetic resonance imaging of the head and a spinal tap are usually done.
- Treatment involves relieving symptoms and sometimes using antiviral drugs.

#### Encephalitis can occur in the following ways:

- A virus directly infects the brain.
- A virus that caused an infection in the past becomes reactivated and directly damages the brain.
- A virus or vaccine triggers a reaction that makes the immune system attack brain tissue (an autoimmune reaction).

Sometimes bacteria cause encephalitis, usually as part of bacterial meningitis (called meningoencephalitis). Protozoa, such as amebas, those that cause toxoplasmosis (in people who have AIDS), and those that causes malaria, can also infect the brain and cause encephalitis. Rarely, encephalitis develops in people who have cancer—a disorder called paraneoplastic encephalitis. What causes it is unknown.

Infections that can directly lead to encephalitis can occur in epidemics or occasionally as isolated cases (sporadically).

#### Epidemic encephalitis:

In the United States, the most common types of epidemic encephalitis are caused by arboviruses. Arboviruses are viruses transmitted to people through the bites of arthropods, usually mosquitoes, fleas, or ticks. (Arbovirus is short for arthropod-borne virus.) The viruses are transmitted to arthropods when arthropods bite infected animals or people. Many species of domestic animals and birds carry these viruses.

Epidemics occur in people only periodically—when the population of mosquitoes or infected animals increases. Epidemics tend to occur when arthropods are biting—for mosquitoes and ticks, usually during warm weather. Infection spreads from arthropod to person, not from person to person.

Many arboviruses can cause encephalitis. The different types of encephalitis that result are usually named for the place the virus was discovered or the animal species that typically carries it.

In the United States, mosquitoes spread several types of encephalitis, including the following:

- La Crosse encephalitis is caused by the La Crosse virus (also called California virus). It is most common in the Midwest but can occur anywhere in the country. This encephalitis accounts for most cases in children. Many cases are mild and undiagnosed. Fewer than 1% of infected people die from it.
- Eastern equine encephalitis occurs predominantly in the eastern United States. It affects mainly young children and people older than 55. In children younger than 1 year, it can cause severe symptoms and permanent nerve or brain damage. Over half of infected people die.
- West Nile encephalitis, once present only in Europe and Africa, first appeared in the New York City area in 1999. It has spread throughout the United States. Several species of birds carry the virus. This encephalitis affects mainly older people. West Nile encephalitis develops in fewer than 1% of people who develop West Nile fever. About 10% of people with West Nile encephalitis die; however, those who have just West Nile fever usually recover fully.
- **St. Louis encephalitis** occurs throughout the United States but particularly in the Southeast (including Florida), Texas, and some Midwestern states. Epidemics once occurred about every 10 years but are now rare.
- Western equine encephalitis can occur throughout the United States but, for unknown reasons, has largely disappeared since 1988. It can affect all age groups but mainly children younger than 1 year.

In other parts of the world, encephalitis is caused by different but related arboviruses. Examples are Venezuelan equine encephalitis and Japanese encephalitis, both spread by mosquitoes.

#### Sporadic encephalitis:

In the United States, sporadic encephalitis is usually caused by herpes simplex virus type 1. Herpes simplex virus causes up to one third of cases of encephalitis. This encephalitis occurs at any time of the year and is fatal if not treated.

Human immunodeficiency virus (HIV) causes a slowly developing brain infection, resulting in HIV-associated encephalopathy (also called HIV-associated or AIDS dementia).

#### Reactivation of a previous infection:

Encephalitis can result from reactivation of a virus, including

- Herpes simplex virus type 1
- Varicella zoster virus (the virus that causes chickenpox)
- JC virus (which causes a usually fatal disorder called progressive multifocal leukoencephalopathy—common among people who have AIDS or other conditions that impair the immune system—see <u>Progressive</u> <u>Multifocal Leukoencephalopathy</u>)
- The virus that causes measles (which, if reactivated, leads to a usually fatal disorder called subacute sclerosing panencephalitis years after measles occurs)

Reactivation can occur long after people have the infection. A reactivated infection can severely damage the brain.

#### Autoimmune encephalitis:

After certain viral infections or vaccines, the body's immune system sometimes attacks the layers of tissue that wrap around nerve fibers (called the myelin sheath) in the brain and spinal cord The attack occurs because proteins in myelin resemble those in the virus. As a result, nerve transmission becomes very slow. The resulting disorder, called acute disseminated encephalomyelitis, resembles multiple sclerosis except that symptoms do not come and go as they do in multiple sclerosis. The viruses most often involved include enteroviruses, Epstein-Barr virus, hepatitis A or B virus, human immunodeficiency virus (HIV), and influenza viruses.

### **Symptoms**

Before symptoms of encephalitis start, people may have digestive symptoms, such as nausea, vomiting, diarrhea, or abdominal pain. Or they may feel as if they are getting a cold or the flu and have cough, fever, a sore throat, a runny nose, swollen lymph nodes, and muscle aches.

#### Symptoms of encephalitis include

- Fever
- Headache
- Personality changes or confusion
- Seizures
- Paralysis or numbness
- Sleepiness that can progress to coma and death

People may vomit and have a stiff neck, but these symptoms tend to be less common and less severe than when caused by meningitis.

Encephalitis due to the herpes simplex virus causes headache, fever, and flu-like symptoms at first. People also have seizures, sometimes accompanied by strange smells, vivid flashbacks, or sudden, intense emotions. As the encephalitis progresses, people become confused, have difficulty speaking and remembering, have repeated seizures, then lapse into coma.

HIV-associated encephalopathy can cause gradual personality changes, problems with coordination, and dementia.

If the spinal cord is affected, parts of the body may feel numb and weak.

Which parts are affected depend on which parts of the spinal cord are affected (see Fig. 1: Where Is the Spinal Cord Damaged? ✓.). People may have difficulty controlling bladder and bowel function. If the infection is severe, people may lose sensation, become paralyzed, and lose control of the bladder and bowels.

## **Diagnosis**

Doctors suspect encephalitis based on symptoms, especially if an epidemic is

#### Did You Know...

 Long after a case of measles or chickenpox, the virus can be reactivated and cause inflammation in the brain.

in progress. Magnetic resonance imaging (MRI) can detect typical abnormalities in the brain, confirming encephalitis. If MRI is not available, computed tomography (CT) may be done. It can help doctors exclude disorders that can cause similar symptoms (such as stroke and brain tumor) and check for disorders that can make doing a spinal tap dangerous.

A spinal tap (lumbar puncture) is done to obtain a sample of cerebrospinal fluid, which flows through the tissues (meninges) that cover the brain and spinal cord. Normally, the spinal fluid contains very few white blood cells. But when the brain and meninges are inflamed, the number of white cells in cerebrospinal fluid increases. To identify the virus causing encephalitis, doctors take samples of blood and cerebrospinal fluid and test them for antibodies to the virus when the person is sick and later when the person is recovering. If the increase in antibodies in cerebrospinal fluid is greater than the increase in the blood, the diagnosis is confirmed. Sometimes techniques are used to grow (culture) viruses in the cerebrospinal fluid so that they can be identified more easily. Enteroviruses can be cultured, but most other viruses cannot.

The polymerase chain reaction (PCR) technique is used to identify many of the viruses that can cause encephalitis. PCR, which produces many copies of a gene, can detect the genetic material of these viruses in a sample of cerebrospinal fluid. Prompt identification of the herpes simplex virus is essential because the encephalitis it causes is destructive and, if untreated, is often fatal. Immediate treatment can help lessen the severity of symptoms and prevent death.

Rarely, a sample of brain tissue is removed and examined under a microscope (biopsy) to determine whether herpes simplex virus or another organism is the cause.

Sometimes, even after extensive testing, no virus, bacteria, or other cause of infection is identified. In such cases, the cause may be autoimmune or paraneoplastic encephalitis because tests cannot always confirm those disorders.

#### **Treatment**

If herpes simplex virus cannot be excluded, the antiviral drug acyclovir is given. Acyclovir is effective against herpes simplex and herpes zoster viruses. Sometimes several antibiotics are also given in case the cause is bacteria. Cytomegalovirus encephalitis can be treated with ganciclovir.

For HIV-associated encephalopathy, a combination of drugs used to treat HIV infection (antiretroviral drugs—see <u>Treatment</u>) helps the immune system function better and delays the progression of the infection and its complications, including dementia.

For other viruses and most other causes, no specific treatment is

available. Treatment usually involves relieving symptoms (such as seizures and fever) and, when necessary, providing life support (for example, with a breathing tube) until the infection subsides—in about 1 to 2 weeks.

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