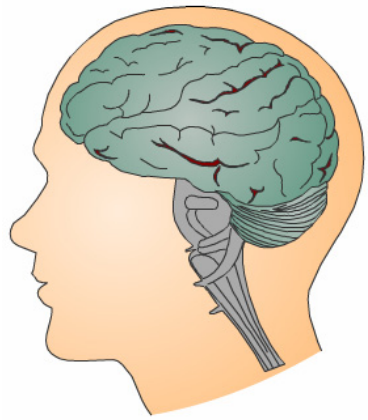
Creutzfeldt-Jakob Disease - CJD

Introduction



CJD stands for Creutzfeldt-Jakob disease. It is a rare brain disorder that leads to death. Each year, it affects about one person in every one million people worldwide.

CJD causes memory problems, behavior changes, vision problems and poor muscle coordination. It progresses quickly. Most patients die within a year.

This reference summary explains Creutzfeldt-Jakob disease. It talks about the causes and symptoms of the disorder. It also covers diagnosis and treatment.

Creutzfeldt-Jakob Disease

CJD is a rare disorder that affects the brain. It usually appears later in life. CJD is a degenerative disorder. This means that CJD impairs the brain's ability to function. Symptoms get worse over time. It leads to dementia and eventual death.

Dementia is the name for a group of symptoms caused by disorders that affect the brain. Memory loss is a common symptom of dementia. Many different diseases can cause dementia.

Many other brain disorders cause symptoms that are similar to those of CJD. But CJD gets worse much more quickly than these other disorders. CJD belongs to a family of human and animal diseases known as TSEs. TSEs stand for transmissible spongiform encephalopathies. "Transmissible" means that the disease can spread. "Spongiform" refers to how the infected brains look. "Encephalopathy" means that it affects the brain. The brain becomes filled with holes and looks spongy under a microscope. CJD is the most common of the known human TSEs. But it is still rare.

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Cattle can get a disease related to CJD called BSE. BSE stands for bovine spongiform encephalopathy. It is commonly known as "mad cow disease."

Causes

CJD and other TSEs seem to be caused by abnormal prions. A prion is a type of protein. They are naturally found in the brain of humans and animals. Normally, prions are harmless. But when they are not shaped correctly, they cause problems in the body. Abnormal prions clump together in the brain. This leads to brain damage. But health care providers do not know exactly how the damage happens.

The risk of getting CJD is low. Most cases of CJD develop for no known reason. These cases are called spontaneous CJD. They may also be called sporadic CJD.

About 5 to 10 percent of all CJD cases are inherited. Inherited means passed down from family member to family member. These cases are caused by a change in the gene that controls how prions are made in the body. CJD that is inherited is also known as hereditary CJD.

Rarely, CJD may be acquired. A small number of people with CJD get it from contact with infected tissue or an infected medical instrument. This usually happens during a medical procedure, such as a skin transplant. It may also happen when people have brain surgery.

There is concern that people can get a form of CJD from eating beef from an infected animal. This is called variant CJD, or vCJD. Research has shown that there may be a connection between mad cow disease and vCJD. But vCJD is very rare.

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Symptoms

CJD is characterized by mental changes that lead to dementia. Symptoms quickly get worse, usually over a few months.

Early signs and symptoms of CJD may include:

- Blurred vision.
- Difficulty speaking.
- Confusion
- Memory loss.
- Personality changes.
- Poor judgment.
- Problems with abstract thinking.
- Sudden mood swings.
- Sudden, jerky movements.
- Trouble sleeping.

Mental symptoms get worse as the disease progresses. These changes can lead to:

- Coma.
- Heart and respiratory failure.
- Pneumonia and other infections.

Most people with CJD die within a year of diagnosis.

Diagnosis

No single test can diagnose CJD. This is because CJD causes symptoms that could be caused by many different conditions. CJD can only be confirmed by examining brain tissue, usually after death. But health care providers can make an accurate diagnosis based on:

- The patient's medical history and symptoms.
- A neurological exam.
- The results of diagnostic tests.

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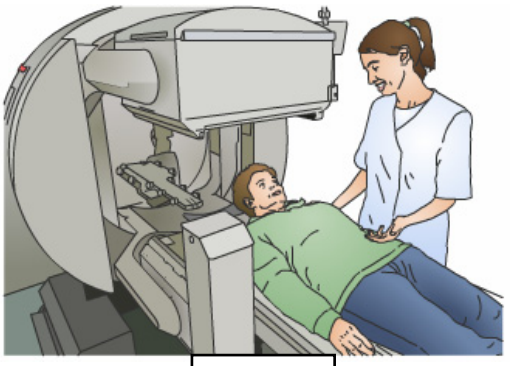
A health care provider will begin by reviewing the patient's medical history. This includes how and when the symptoms began. It also includes talking about any health issues that may help identify another cause of the symptoms, such as recent head trauma. A health care provider will also ask about the patient's family medical history. In some cases, CJD may be inherited.

When diagnosing a condition that causes problems with memory, a health care provider may want to interview friends, family and other caregivers. These people are able to share concerns about the changes they have witnessed that the patient might be unaware of. The health care provider will also perform a physical exam. This will include a neurological exam. It can reveal symptoms, including:

- Memory problems.
- Muscle weakness or spasms.
- Abnormal or slowed reflexes.
- Problems with coordination.

Diagnostic tests may be used to help diagnose CJD. These tests can also help rule out other causes of the patient's symptoms. Brain scans and other tests can identify strokes, tumors or other problems that may cause symptoms of CJD. These include:

- Magnetic resonance imaging, or MRI.
- Electroencephalogram, or EEG.



MRI

An MRI is used to make detailed pictures of areas inside your body. It uses strong magnets to create images of the inside of the body. An EEG detects and records patterns of electrical activity in the brain. To do this, electrodes are placed on the scalp. An EEG checks for abnormal patterns in brain wave activity.

A spinal tap may also be done. This procedure collects a small sample of cerebrospinal fluid, or CSF. The sample is then sent to the lab for testing. In patients with CJD or vCJD, a particular protein is often found in the sample.

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Rarely, a brain biopsy may be done. A biopsy is the removal of tissues for examination by a pathologist.

Treatment

There is no treatment that can cure or control CJD. Current treatments focus on controlling the symptoms of the disease. They also help the patient remain comfortable as the disease progresses. Most people with CJD die about seven months after diagnosis. But some people may live up to one or two years after being diagnosed with CJD.

Research is being done to find treatments for CJD. A variety of drugs are currently being tested.

Summary

CJD stands for Creutzfeldt-Jakob disease. It is a rare brain disorder that leads to death. Each year, it affects about one person in every one million people worldwide. CJD causes memory problems, behavior changes, vision problems and poor muscle coordination.

CJD seems to be caused by abnormal prions that clump together in the brain. This leads to brain damage. CJD may be:

- Sporadic, which happens for no known reason.
- Hereditary, which runs in families.
- Acquired, which happens from contact with infected tissue, usually during a medical procedure.

There is concern that people can get a form of CJD from eating beef from an infected animal. This is called variant CJD, or vCJD. Research has shown that there may be a connection between mad cow disease and vCJD. But vCJD is very rare.

CJD gets worse over time. It leads to dementia and, eventually, death. Most patients die within a year of diagnosis. Current treatments focus on controlling pain and other symptoms.

Research is being done to find better treatments for CJD. A variety of drugs are currently being tested.

Last reviewed: 01/02/2019