

Disease Progression

The different subtypes of Creutzfeldt-Jakob disease (CJD) differ somewhat in typical symptoms and progression.

Sporadic CJD

This is often reported as having "come out of the blue." Early symptoms may look like depression but quickly progress (within a few weeks) to the confusion and memory problems commonly seen in dementia. Neurological difficulties, such as loss of coordination and balance (called "progressive cerebellar ataxia") together with progressive blindness usually follow, but are occasionally the first signs of CJD. First symptoms usually show up in a person's 60s.

As the disease progresses, people with sporadic CJD become increasingly confused and unable to walk, communicate, be aware of their surroundings or care for themselves. Brief jerking movements of the muscles (called myoclonus) and difficulty swallowing occur in most cases. People usually lose the ability to speak or move, which makes them susceptible to potentially fatal pneumonia. The illness is usually distressingly rapid. Three-quarters of people with sporadic CJD die within six months of diagnosis, while others die within just a few weeks. Rarely, the illness may last for a year or more.

Familial CJD

The symptoms of genetic/familial CJD vary between different people, depending partly on the type of gene mutation responsible. In some cases the illness is very like that of sporadic CJD in type, duration and progression, while in others it is a more slowly developing dementia which progresses over a few years.

Acquired CJD

Iatrogenic CJD

The symptoms of iatrogenic CJD vary according to the cause. The signs and symptoms following human dura mater grafts are generally similar to those of sporadic CJD. Human growth hormone cases follow a different course, with dementia and a progressive loss of coordination and balance developing slowly over many months.

Variant CJD

vCJD usually affects younger people and begins with a range of mainly emotional or behavioral disturbances, such as social withdrawal, depression, excessive sleepiness, anxiety or agitation. Sometimes, delusions (abnormal beliefs) and hallucinations (abnormal experiences, including seeing or hearing things which aren't there) occur. In addition, patients may have unpleasant sensations, pain or numbness in the limbs or other parts of the body.

These initial features may be difficult to distinguish from primarily psychiatric or psychological problems. Within months of the psychiatric symptoms, neurological symptoms develop and usually include poor balance and clumsiness. Abnormal movements such as the restless, dance-like movements of chorea or the rigid posture of dystonia may be apparent. They may stop speaking. Typically, jerky muscle movements (myoclonus) appear in the later stages, and forgetfulness and confusion develop into dementia.

People with vCJD gradually become unable to do things for themselves, and their increasing mental impairment leads to dependence on caregivers for ordinary daily activities. They may develop eating and swallowing difficulties and require artificial feeding. The average life expectancy following diagnosis is just over a year, and the longest recorded duration a little more than three years.

Other signs and symptoms of vCJD include:

- A sense that the skin feels sticky

- Sensations of cold or pain
- Muscle paralysis
- Tremors