Creutzfeldt - Jakob Disease

Creutzfeldt-Jakob disease (CJD) is a rare degenerative disease that affects brain tissue in humans. When people first start to show symptoms they usually present with confusion or disorientation and problems with walking. While death can occur up to two years after a person shows the first symptoms, most people die within six months. There is no treatment or cure.

There are two different types of CJD - classical CJD and variant CJD.

Classical CJD is a rapidly progressive and fatal disease of the brain that occurs in Australia in about one per million people per year. There are three forms of classical CJD:

- Sporadic CJD is the most common form of CJD, and usually occurs in people over the age of 40 years, most often between 50 to 70 years. The cause of sporadic CJD is unknown.
- Health care acquired CJD has occurred worldwide as a result of a few specific medical treatments mostly involving brain tissue. Once CJD was identified as a hazard, the use of these treatments was stopped. Ten deaths have occurred from health care acquired CJD in Australia, but none since 1991.
- Familial (genetic) CJD is extremely rare. It accounts for 5-15% of classical CDJ cases and there is usually a family history of the illness.

Variant CJD was first recognised in 1996 in the United Kingdom (UK). Variant CJD is thought to be linked to eating meat from animals infected with bovine spongiform encephalopathy (BSE), better known as 'mad cow' disease. BSE has not been found in Australian livestock. There has been no reported human case of variant CJD in Australia to date.

Signs and Symptoms:

Early symptoms include changes in personality and behaviour, decline in thinking ability, visual abnormalities, muscle weakness and loss of control over movement. Confusion, speech abnormalities, agitation and hallucinations may occur. Eventually the disease progresses to dementia, coma and death.

CJD is difficult to diagnose. The early symptoms are very general and there are no actual tests to diagnose the disease until symptoms are well advanced. As the disease progresses, many investigations are necessary to rule out the possibility of other treatable diseases. Examination of brain tissue after death is the only way to confirm CJD.

It is essential to remember that most people with these symptoms do NOT have CJD, and are experiencing the symptoms due to other causes.

Treatment:

At the moment, there is no effective treatment for CJD. Current treatment is aimed at relieving the symptoms and making the person as comfortable as possible.

Prevention:

In Australia, CJD is a nationally notifiable disease. CJD is not spread by person to person, household, social or sexual contact. There is no need for special precautions in the home or community to prevent the spread of the disease.

Australia has banned beef imports from the UK since 1998, and has also banned imports from other BSE affected countries. No cases of BSE have been reported in Australian livestock and a certification system exists to ensure all beef products entering Australia are free of BSE.

For those who have travelled to or lived in the UK between 1980 and 1996, the risk of contracting variant CJD is considered to be very low, even among those who ate beef. Despite this very low risk, people who spent six months or more in the UK between 1980 and 1996, as well as people who received a blood transfusion in the UK from 1980 onwards, are declined as blood donors.

Stronger guidelines have also been established for the use of organ transplants and for the sterilisation of surgical equipment to reduce the risk of CJD acquired through organ donation or through surgical equipment.

Health outcome:

Although the disease is very rare it is usually fatal.

Help and assistance:

For further information please contact your local doctor, nearest public health unit or 13HEALTH (13 43 25 84).

References

Australian Government Department of Health and Ageing (http://www.health.gov.au/internet/main/publishing.nsf/Content/icg-guidelines-index.htm) (2007). Infection Control Guidelines - Creutzfeldt-Jakob Disease.

URL: http://conditions.health.qld.gov.au/HealthCondition/condition/6/48/277/creutzfeldt-jakob-disease
Version number: 5
Date published: 20/12/2017
Date generated: 25/10/2021

Heymann D (Ed) (2008). Control of Communicable Diseases Manual, 19th edition. Washington, DC: American Public Health Association, pp 94-99.

Mandell G, Bennett J & Dolin R (2009). Principles and Practices of Infectious Diseases, 7th edition. Elsevier Inc US.

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Creutzfeldt-Jakob disease (https://www.healthdirect.gov.au/creutzfeldt-jakob-disease)

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