FACT*SHEET*
NSW HEALTH

CREUTZFELD-JAKOB DISEASE

WHAT IS CREUTZFELD-JAKOB DISEASE?

Creutzfeldt-Jakob disease (CJD) is a rare and fatal brain disease in humans. It is a type of disease known as a transmissible spongiform encephalopathy (TSE) because it causes characteristic spongy breakdown of the brain and it can be transmitted. Other animals—such as sheep, cows and cats—can also develop TSEs.

THE FOUR MAIN TYPES OF CJD.

Sporadic (classical) CJD
This is the most common form, responsible for 85 per cent of cases. The cause is unknown. It mainly affects people aged over 50 years.

Familial CJD
This is an inherited form of the disease with a younger age of onset. It causes 10–15 per cent of cases of CJD.

Iatrogenic CJD
This occurs through the inadvertent use of infectious material in medical procedures.

Variant CJD
This is a newly-recognised type that was first discovered in 1996 in the United Kingdom. It is caused by the same infectious agent that causes 'mad cow disease' (BSE or Bovine Spongiform Encephalopathy) that has affected cattle in the United Kingdom and other parts of Europe. It is different to sporadic CJD because it usually affects younger people, who have a longer duration of illness. It also has slightly different clinical features and can be distinguished from sporadic CJD by postmortem laboratory examination of brain tissue (that is, after the person has died). Evidence of the infection can also be found in lymph tissue, such as tonsils.

WHAT CAUSES CJD?

It is thought that an infectious agent, known as a prion, causes the damage to the brain. Prions are different to other infectious agents (such as bacteria and viruses) because they are made from a protein that is normally present in all cells. It is believed that the prion causes normal cell proteins to change into abnormal cell proteins, and that these build up in the brain, causing damage.

In inherited CJD the genes that tell the body how to make the protein may be faulty. In iatrogenic CJD the abnormal protein comes from contaminated tissue or instruments. Variant CJD is thought to occur when the prion is ingested in contaminated beef or beef products. It is unclear if other risk factors or predisposing factors are needed to enable the prion to cause disease. In sporadic CJD no one knows how the abnormal protein arises.

HOW COMMON IS CJD?

CJD is rare. It occurs worldwide at a rate of 0.5 to 1 case per million per year. Overall, Australia has about the same case rate as other countries. Because of the relationship with Bovine Spongiform Encephalopathy, most cases of variant CJD have occurred in the United Kingdom (a total of 85 cases as of November 2000). An additional three cases have been reported in France and one in Ireland. Because CJD can have an incubation period of many years, it is unclear how many new cases of variant CJD will occur. *As of February 2001 no cases of variant CJD or BSE have been reported in Australia.*

WHAT ARE THE SYMPTOMS OF CJD?

CJD causes progressive symptoms of difficulty with coordination, muscle jerks and memory loss with eventual dementia. Variant CJD can also cause mood disturbance and altered sensations. CJD is fatal, usually within a year of onset of symptoms. At present there is no cure. Treatments for symptoms such as pain and muscle jerks are very important in caring for people with CJD.

HOW IS CJD DIAGNOSED?

A definite diagnosis can only be made by examining brain tissue after death. There are however other tests such as CAT scans, MRI scans and EEGs that can be highly suggestive of the diagnosis and are used to rule out other diagnoses. People with variant CJD may have the prion protein present in their tonsils and other tissues.

CAN YOU CATCH CJD?

In Australia, iatrogenic CJD has been caused by the use of human growth hormone taken from the pituitary glands of cadavers that are infected with CJD. Currently, as there is no Bovine Spongiform Encephalopathy in Australian cattle, no risk is posed by eating Australian beef or beef products. Beef and beef products from countries with infected cattle have been banned from import to Australia.

There is no evidence that sporadic CJD is spread by blood transfusion. There is a theoretical possibility that variant CJD could be transmitted by blood transfusion, although this has never been reported. As a precautionary measure, people who have spent six months or more in Great Britain between 1980 and 1996 cannot donate blood. This is because beef and beef products consumed in Great Britain during this time could have come from cows harbouring BSE infection. Regular surveillance for BSE in cattle is carried out routinely in Australia.

There is no evidence that CJD can be transmitted by normal social contact.

For further information contact your doctor, community health care centre, or your nearest public health unit.