

## Creutzfeldt-Jakob disease (CJD)

### Background Information

CJD is a very rare, fatal neurodegenerative disease, which is thought to be caused by the build up of an abnormal form of the naturally occurring 'prion' protein in the brain.

There are four types of CJD (variant, sporadic, inherited and iatrogenic), of which only variant CJD (vCJD) is an occupationally acquired disease.

CJD belongs to a group of Transmissible Spongiform Encephalopathies (TSEs) which affect humans and animals.

### Organism:

Bovine Spongiform Encephalopathy (BSE) agent

### Incidence and Transmission:

The first reported case of vCJD was in 1996 and since then there have been almost 200 deaths in the UK.

vCJD is strongly linked to exposure to – and in particular consumption of – beef or beef products from cattle suffering from BSE or 'mad cow disease'. The first reported case of BSE in cattle in the UK was in 1986. BSE was spread via feed made from rendered material from infected cattle or sheep, a process which has been banned since 1988.

There has never been a reported case of disease following occupational exposure (despite more than 184 600 cases in cattle in the UK), however it remains a theoretical risk. It is thought the most likely route of occupational infection would be via broken skin and it is thought unlikely that the disease would be transmitted via aerosols.

There is no evidence of spread from person to person, but this remains theoretically possible.

### Occupations and processes where vCJD may present a risk

Occupational exposure to vCJD may occur in those who:

- are in contact with infected animals; or
- work with materials or products from infected animals, in particular brain, spinal cord and lymphatic tissue.

Occupations where there may be a risk of occupationally acquired vCJD include:

- farmers;
- veterinary surgeons;

- abattoir workers, meat and bone processing plant workers and butchers;
- zoo and circus workers;
- incinerator operators;
- cleaning and waste disposal workers and landfill site workers;
- maintenance engineers (abattoirs, rendering plants and incinerators); and
- healthcare workers.

### **Clinical Information**

The incubation period for vCJD is unknown, but is generally several years.

The symptoms of vCJD include psychiatric problems and uncontrolled movements. There is no treatment or cure and this disease is always fatal, with an average life expectancy of around one year from diagnosis.

Anyone with these symptoms should seek medical attention. There is no laboratory test for vCJD and diagnosis is based on clinical symptoms and is confirmed by post mortem microscopic examination of brain tissue.

### **Control**

The following control measures reduce the risk of infection:

- There are UK controls for the handling of nervous and lymphatic tissue (Specified Risk Material) from animals.
- There are strict UK controls governing the preparation of beef and beef products for consumption and the handling and disposal of infected cattle ([UK BSE controls](#)).
- Full personal protective equipment, including a fit tested respirator must be worn when handling infected material.
- Suitable cleaning procedures should be used – the BSE agent is resistant to most disinfectants.
- Good occupational hygiene practices should be followed, especially washing with warm water and soap.
- Cuts and abrasions should be covered with waterproof plasters.

### **Further Information**

[HSE – BSE Occupational Guidance](#)

[Public Health England – CJD](#)

[World Organisation for Animal Health – BSE Information](#)

[NHS Choices – vCJD](#)