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Cumbria County Council

CREUTZFELDT-JAKOB DISEASE

Community Infection Prevention and Control Guidance for Health and Social Care

Creutzfeldt-Jakob Disease

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CREUTZFELDT-JAKOB DISEASE

This guidance document has been adopted as a policy document by:

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1. Introduction

Creutzfeldt-Jakob Disease (CJD) is one of a group of diseases called Transmissible Spongiform Encephalopathies (TSEs) which can occur in people or animals. The transmissible agent is an abnormal protein known as a prion. TSEs are characterised by degeneration of the nervous system and are invariably fatal.

CJD has a long incubation period and may not cause symptoms for many years. Clinical features vary depending on the regions of the brain affected, but all service users experience a very rapid deterioration following onset of symptoms. There are no simple non-invasive tests available to diagnose CJD before symptoms develop, diagnosis can only be confirmed on the death of a service user by a brain biopsy.

In this guidance, the term CJD encompasses sporadic CJD, variant CJD (vCJD), familial CJD, and other TSEs. There are several types of CJD:

- **Sporadic**: commonest form caused by a mutant gene. Usual age of onset is late middle age. Most service users present with rapidly progressive dementia with focal neurological signs including ataxia, myoclonus, visual disturbances and rigidity. Death occurs within 4-6 months of clinical onset.
- **Familial:** approximately 15% of cases are inherited and caused by a gene mutation.
- **Iatrogenic**: about 1% are transmitted by medical or surgical procedures including pituitary hormone injections, dura mater grafts, and rarely by neurosurgical instruments. The incubation period can range from 1-2 years for neurological routes of transmission and up to 30 years in some pituitary hormone recipients.
- **Variant CJD** (vCJD): thought to be as a result of eating contaminated bovine food products (same agent responsible for BSE in cattle). Whilst rare, there has been a gradual increase in numbers of people being diagnosed. Tends to affect young adults, with the clinical illness lasting an average of 14 months. Symptoms may include both psychiatric and sensory abnormalities, which are followed by ataxia, myoclonus and other movement disorders and dementia.

2. Transmission

How TSE's are transmitted is uncertain, but there is no evidence that they are spread from person-to-person by close contact. It is, however, known that transmission of Sporadic CJD can be associated with medical intervention,

e.g., administration of hormones prepared from human pituitary glands, dura mater preparations, corneal grafts and recently from blood transfusions. CJD/ vCJD has also been reported following brain surgery due to inadequately decontaminated instruments (prion proteins are resistant to decontamination processes).

The Advisory Committee on Dangerous Pathogens has suggested that in people with sporadic CJD, certain tissues have high, medium or low infectivity. There is evidence that the distribution of the abnormal prion protein in tissues is more widespread in the body in service users with vCJD, than in service users with sporadic CJD.

Tissue infectivity of CJD and vCJD

Tissue	Assumed level of infectivity	
	CJD other than vCJD	vCJD
Brain	High	High
Spinal cord	High	High
Cranial nerves, specifically the entire optic nerve and only the intracranial components of the other cranial nerves	High	High
Cranial ganglia	High	High
Posterior eye, specifically the posterior hyaloid face, retina, retinal pigment epithelium, choroid, sub-retinal fluid, optic nerve	High	High
Pituitary gland	High	High
Spinal ganglia	Medium	Medium
Olfactory epithelium	Medium	Medium
Dura mater	Low	Low
Tonsil	Low	Medium
Lymph nodes and other organised lymphoid tissues containing follicular structures	Low	Medium
Gut-associated lymphoid tissue	Low	Medium
Appendix	Low	Medium
Adrenal gland	Low	Medium
Spleen	Low	Medium
Thymus	Low	Medium
Anterior eye and cornea	Low	Low
Peripheral nerve	Low	Low
Skeletal muscle	Low	Low
Dental pulp	Low	Low
Gingival tissue	Low	Low
Blood and bone marrow	Low	Low
CSF	Low	Low
Placenta	Low	Low
Urine	Low	Low
Other tissues	Low	Low

3. Risk groups

When considering measures to prevent transmission to service users or staff in a health and social care setting, it is useful to make a distinction between:

- symptomatic service users, i.e., those who fulfil the diagnostic criteria for definite, probable or possible CJD or vCJD; and
- service users 'at increased risk', i.e., those with no clinical symptoms, but who are 'at increased risk' of developing CJD or vCJD, because of their family or medical history.

It is the responsibility of the clinician to ensure that an assessment to determine risk is undertaken using the table below as guidance.

Risk groups	
Symptomatic service users	• Individuals who fulfil the diagnostic criteria for definite, probable or possible CJD or vCJD • Individuals with neurological disease of unknown aetiology, who do not fit the criteria for possible CJD or vCJD but where the diagnosis of CJD is being actively considered
Asymptomatic service users 'at risk' from genetic forms of CJD	• Individuals who have a blood relative known to have a genetic mutation indicative of genetic CJD • Individuals who have been shown by specific genetic testing to be at significant risk of developing CJD
Asymptomatic service users identified as 'at increased risk' of CJD/vCJD through iatrogenic exposure	• Recipients of hormone derived from human pituitary glands, e.g., growth hormone, gonadotrophin • Individuals who have received a graft of dura mater (people who underwent neurosurgical procedures or operations for a tumour or cyst of the spine before August 1992 may have received a dura mater graft and should be treated as at risk unless evidence can be provided that dura mater was not used) • Individuals who have been contacted as potentially at risk for public health

4. Care of a service user with CJD

Normal social or routine clinical contact with a service user with CJD or related disease does **not** present a risk to healthcare staff, relatives or the community. Isolation is not necessary and they can be cared for at home or in a health and social care setting. No special measures over and above standard infection control precautions are required for caring for CJD or vCJD service users in a community setting, as it is **unlikely** that a procedure will be undertaken that involves contact with high or medium risk tissues.

Although cases of CJD/vCJD have been reported in healthcare staff, there have been no confirmed cases linked to occupational exposure.

The following advice is for the care of service users who are known, suspected or at risk of developing CJD or related disorders.

Communication	Your local Community Infection Prevention and Control team or Public Health England team should be contacted in order to give appropriate advice
Type of isolation	Isolation is not required. A service user may be cared for in their own home or in a health and social care setting and can socialise and take part in normal activities
Main infection source	The main potential source of infection is from high risk tissues, especially brain, spinal cord, eye and cerebrospinal fluid (CSF) in sporadic CJD, contact with high risk tissues is unlikely in a community setting. There is no evidence of infectivity in saliva, body excretions or excreta. As the infectivity of other tissues in vCJD is less well understood, standard precautions should be adhered to including covering cuts and abrasions with a waterproof dressing
Pathology specimens	All specimens from a service user with a definite, probable or possible diagnosis of CJD, must be labelled as 'infection risk'. Pathology specimens should only to be taken if absolutely essential, and after prior consultation with your local Community Infection Prevention and Control or Public Health England team and the pathology laboratory
Personal protective equipment	Disposable apron and gloves should be worn when performing any procedure which involves handling tissues, blood or body fluids and face protection if splashing is anticipated
Disposal of faeces/urine	No specific precautions are required. Service users may use the toilet provided good personal habits are maintained. If a commode is used, the commode and pan should be designated for that service user

Disposal of infectious waste	Infectious waste should be disposed of as per local policy
Cutlery and crockery	No specific precautions. Disposable items are not required
Medical equipment	Single use equipment should be used, where possible, if in contact with body fluids and disposed of as infectious waste. Medical equipment in contact with intact skin should be decontaminated with detergent and warm water or an alcohol wipe after use
Linen	No special requirements, linen and clothing should be laundered as usual

5. Clinical and surgical procedures

The advice of your local Community Infection Prevention and Control or Public Health England team must always be sought before any clinical or surgical procedure on known, suspected or at risk individuals.

6. Spillages of blood and body fluids

Spillages in a community setting will be of low risk material, e.g., blood and urine. Standard precautions should be followed when dealing with spillages of blood and body fluids from service users 'with' or 'at increased risk' of CJD/vCJD. A chlorine-based disinfectant solution at a dilution of 10,000 parts per million (ppm) should be used for the disinfection of a blood spillage (see Decontamination, Cleaning and Disinfection Guidance). A chlorine-based disinfectant solution at a dilution of 1,000 ppm should be used for the disinfection of a body fluid spillage.

It should be noted that chlorine-based disinfectant (hypochlorite) solutions will not deactivate TSE agents.

7. Accidental inoculation injury

Any accident involving 'sharps' or contamination of abrasions with blood or body fluids should be dealt with in accordance with the Sharps Management and Inoculation Injuries Guidance and reported immediately to the Occupational Health Department/GP practice /A&E department, who will discuss the case with a Consultant Microbiologist.

8. Contact lenses and ophthalmic devices

There have been no known cases of iatrogenic transmission of CJD/vCJD resulting from diagnostic examination or contact lens wear. Although contact with the corneas is considered as low risk in terms of iatrogenic transmission, further advice can be obtained from the Department of Health's 'Guidance from the ACDP TSE Risk Management Subgroup'.

The use of single-use instruments or contact lenses is recommended for use on those designated at increased risk of CJD or vCJD.

9. Death of a service user

Inform your local Community Infection Prevention and Control or Public Health England team. Relatives of the deceased may wish to view or have some final contact with the body. Such viewing and possible contact such as kissing need not be discouraged.

The undertaker must be informed of the infection status. It is recommended that the deceased person's body is placed in a cadaver bag prior to transportation to the undertakers or mortuary.

Under no circumstances must any tissue or organs be used for donation.

10. Undertakers

Concern about possible unknown CJD cases does not warrant a level of precaution for undertakers handling intact bodies other than those used generally for all work of this nature. Dressing and cosmetic work on deceased service users from a risk group may be undertaken, if the usual precautions routinely used when dealing with the deceased are observed.

Where the diagnosis of CJD is known or suspected, it is advisable to avoid embalming procedures.

11. Funerals and cremations

There is no need for extra precautions to be taken for either burial or cremation. Relatives, friends or carers of the deceased may wish to view or have some final contact with the deceased. Such viewing and possible superficial contact, such as kissing, or touching, need not be discouraged.

12. Dental treatment

The risks of transmission of infection from dental instruments are thought to be very low provided satisfactory standards of infection control and decontamination are maintained. There is no reason why any service user 'with' or ‘at increased risk’ of CJD or vCJD should be refused routine dental treatment. Such people can be treated in the same way as any member of the general public.

Service users requiring dental care must inform their dental practitioner before treatment. Their status is then recorded in the notes.

Dental instruments used on service users 'with' or ‘at increased risk’ of CJD or vCJD can be handled in the same way as those used in any other low risk surgery, i.e., these instruments can be reprocessed according to best practice and returned to use.

Advice on the decontamination of dental instruments can be found in the Department of Health guidance HTM 01-05 ‘Decontamination in primary care dental practices’.

Single use instruments should be disposed of after one use and never reprocessed under any circumstances.

The service users ‘at risk’ status must be included in any referrals for surgery as head and neck surgery may involve contact with tissues of high or medium infectivity, for which special infection control precautions will be required.

13. Additional IPC resources

The North Yorkshire and York Community Infection Prevention and Control (IPC) team have produced a wide range of innovative educational and other IPC resources including support for decontamination, cleaning and disinfection, e.g., National colour coding scheme posters. These resources are designed to assist your organisation in achieving compliance with the Health and Social Care Act 2008 and CQC requirements. Further information on these high quality evidence-based resources is available at www.infectionpreventioncontrol.co.uk

14. References

Department of Health (2013) *Minimise transmission risk of CJD and vCJD in healthcare settings*

Department of Health (2013) *Decontamination Health Technical Memorandum 01-05: Decontamination in primary care dental practices*

Department of Health (Updated 2013) *Guidance: Minimise transmission risk of CJD and vCJD in healthcare settings* https://www.gov.uk/government/publications/guidance-from-the-acdp-tse-risk-management-subgroup-formerly-tse-working-group [Accessed 29 April 2015]

Health and Safety Executive (2005) *Controlling the risks of infection at work from human remains. A guide for those involved in funeral services (including embalmers) and those involved in exhumation*