GP 04

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CREUTZFELDT-JAKOB DISEASE

Community Infection Prevention and Control Guidance for General Practice

(also suitable for adoption by other healthcare providers, e.g. Dental Practice, Podiatry)

Creutzfeldt-Jakob disease

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GP 04

CREUTZFELDT-JAKOB DISEASE

Please note that the internet version is the only version that is maintained. Any printed copies should, therefore, be viewed as 'uncontrolled' and as such, may not necessarily contain the latest updates and amendments.

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

Organisation: Brigstock Family Practice

Signed: 

Job Title: Registered Manager

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If your organisation would like to exclude or include any additional points to this document, please include below. Please note, the Community IPC Team cannot endorse or be held responsible for any addendums.

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CREUTZFELDT-JAKOB DISEASE

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CREUTZFELDT-JAKOB DISEASE

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 patients 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 patient 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 patients 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 patients with vCJD, than in patients with sporadic CJD.

Tissue infectivity of CJD and vCJD

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

3. Risk groups

When considering measures to prevent transmission to patients or staff, it is useful to make a distinction between:

- Symptomatic patients, i.e. those who fulfil the diagnostic criteria for definite, probable or possible CJD or vCJD; and
- Patients '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 patients	• 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 patients '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 patients 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 and asked to follow public health precautions

4. Care of a patient with CJD

Normal social or routine clinical contact with a patient 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 patients 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 patients who are known, suspected or at risk of developing CJD or related disorders.

Communication	Your local Community Infection Prevention and Control or Public Health England Team should be contacted in order to give appropriate advice
Type of isolation	Isolation is not required. A patient 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 patient 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. Patients may use the toilet provided good personal habits are maintained
Disposal of	**Clinical waste should be disposed of as per local**

Clinical waste	**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 detergent or disinfectant wipes 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 should be dealt with as per the 'Decontamination, cleaning and disinfection guidance'.

7. Inoculation injury and blood or body fluid splashes

Any incident involving used sharps, splashes into the eyes, mucous membranes 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. Referral or transfer to other health and social care provider

It is a requirement of *The Health and Social Care Act 2008 Code of Practice on the prevention and control of infections and related guidance* (The Code of Practice), that primary medical care practices provide accurate information on the infection status of a service user when transferring them to another health or social care provider.

Utilising the Inter-Health and Social Care Infection Control Transfer Form ensures (see Appendix 1):

- Accurate information is communicated in an appropriate and timely manner
- This information facilitates the provision of optimum care, minimising the risk of inappropriate management and further transmission of infection
- Where possible, information accompanies the patient

The completed form should be supplied to the receiving provider and a copy kept by the transferring provider in the patient's records.

The patients '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.

10. Death of a patient

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.

11. Infection Prevention and Control resources, education and training

The Community Infection Prevention and Control (IPC) Team have produced a wide range of innovative educational and IPC resources designed to assist

your Practice in achieving compliance with the *Health and Social Care Act 2008* and CQC registration requirements.

These resources are either free to download from the website or available at a minimal cost covering administration and printing:

- Over 20 IPC Guidance documents (Policies) for General Practice
- 'Preventing Infection Workbook for General Practice'
- 'IPC CQC Inspection Preparation Pack for General Practice'
- IPC audit tools, posters, leaflets and factsheets
- 'IPC Advice Bulletin for GP Practice Staff'

In addition, we hold educational study events in North Yorkshire and can arrange bespoke training packages and 'Mock IPC CQC Inspections'. Prices vary depending on your requirements and location.

Further information on these high quality evidence-based resources is available at www.infectionpreventioncontrol.co.uk.

12. References

Department of Health (2012 updated October 2015) *Minimise transmission risk of CJD and vCJD in healthcare settings* www.gov.uk/government/publications/guidance-from-the-acdp-tse-risk-management-subgroup-formerly-tse-working-group

Department of Health (2003 revised and updated February 2015) *Transmissible Spongiform Encephalopathy Agents: Safe Working and the Prevention of Infection: Part 4 Infection Prevention and Control of CJD and Variant CJD in Healthcare and Community Settings* www.gov.uk/government/uploads/system/uploads/attachment_data/file/427854/Infection_controlv3.0.pdf

World Health Organisation *Creutzfeldt Jakob Disease* www.who.int/topics/creutzfeldtjakob_syndrome/en/

13. Appendices

Appendix 1: Inter-Health and Social Care Infection Control Transfer Form

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Inter-Health and Social Care Infection Control Transfer Form

The *Health and Social Care Act 2008: Code of Practice on the prevention and control of Infection and related guidance* (Department of Health 2015), states that "suitable accurate information on infections be provided to any person concerned with providing further support or nursing/medical care in a timely fashion". This form has been developed to help you share information with other health and social care providers. The form should accompany the patient and, where possible, a copy filed in the patient's notes.

Patient Name: ..... Address: ..... NHS number: ..... Date of birth: ..... Patient's current location:	GP Name and contact details:

Receiving facility, e.g., hospital ward, hospice: .....

If transferred by ambulance, the service has been notified: Yes ☐ N/A ☐

Is the patient an infection risk:
Please tick most appropriate box and give details of the confirmed or suspected organism

☐ Confirmed risk Organisms: .....

☐ Suspected risk Organisms: .....

☐ No known risk

Patient exposed to others with infection, e.g., D&V, Influenza: Yes ☐ No ☐ Unaware ☐

If yes, please state: .....

If the patient has a diarrhoeal illness, please indicate bowel history for last week, if known, (based on Bristol Stool Form Scale):

.....

Is diarrhoea thought to be of an infectious nature? Yes ☐ No ☐ Unknown ☐

Relevant specimen results if available

Specimen:				
Date:				
Result:				

Treatment information:

Is the patient aware of their diagnosis/risk of infection? Yes ☐ No ☐

Does the patient require isolation? Yes ☐ No ☐

If the patient requires isolation, phone the receiving facility in advance: Actioned ☐ N/A ☐

Additional information:

Name of staff member completing form: .....

Print name: .....

Contact No: ..... Date .....

Community Infection Prevention and Control, Harrogate and District NHS Foundation Trust April 2017
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