Creutzfeldt-Jakob Disease (CJD) Procedure

(IPC Manual)
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1. **INTRODUCTION**

Transmissible spongiform encephalopathies (TSEs), also known as prion diseases are a group of rare, degenerative brain diseases that affect humans and some animals. Prion diseases exist in different forms, all of which are progressive, currently untreatable and ultimately fatal.

Sporadic Creutzfeldt-Jakob disease (CJD) is one of the TSEs and was first recognised in the 1920’s as a fatal disease affecting the brain. In 1996 a variant new strain of the disease (vCJD) was identified. Further research established the relationship between exposure to Bovine Spongiform Encephalopathy (BSE) found within cattle and vCJD.

At present there is no evidence to suggest that CJD or vCJD is transmitted from person to person via close contact. However, it has been noted that sporadic CJD has been associated with a variety of medical interventions including: administration of hormones prepared for the pituitary gland, corneal grafts, blood transfusions and following neuro-surgery with contaminated instruments.

2. **PROCEDURE**

2.1 **Infection risk**

The highest potential risk is from exposure to high infectivity tissues through direct inoculation, for example as a result of sharps injury, puncture wound or contamination of broken skin, and mucous membrane exposure. Healthcare personnel who work with patients with definite/probable/possible CJD/vCJD, or with potentially infected tissues should be appropriately informed about the nature of the risk and relevant safety procedures. Compliance with standard infection prevention and control (IPC) precautions will help to minimise risks from occupational exposure.

When considering measures to prevent transmission in a healthcare setting, it is useful to distinguish between **symptomatic** patients, who fulfill the diagnostic criteria for definite, probable or possible CJD or vCJD and patients with no clinical symptoms, who are “at increased risk” of developing one of these diseases because of their medical or family history (appendix A).

There is no evidence that normal social or routine clinical contact with a CJD/vCJD patient presents a risk to healthcare workers, relatives and others. Isolation of patients with CJD/vCJD is not necessary, and they can be nursed in an open ward using standard IPC precautions in line with those used for all patients.

2.2 **Sample taking and other invasive medical procedures**

When taking samples or performing other invasive procedures, the possible infectivity of the tissue(s) involved must be considered, and if necessary suitable precautions taken (appendix B).
It is important to ensure that only suitably trained staff, who are aware of the hazards, carry out invasive procedures that may lead to contact with medium or high risk tissue.

Body secretions, fluids (including saliva, blood and cerebrospinal fluid (CSF)) and excreta are all low risk for CJD/vCJD. It is therefore likely that the majority of samples taken or procedures performed will be low risk. Contact with small volumes of blood (including inoculation injury) is considered low risk, though it is known that transfusion of large volumes of blood and blood components may lead to vCJD transmission. Blood and body fluid samples from patients with or “at increased risk” of CJD/vCJD should be handled using standard infection prevention and control precautions as for any other patient including:

- Use of disposable gloves, aprons and eye protection where body fluid exposure/splash anticipated
- Use of medical devices that incorporate safety-engineered protection mechanisms such as needle safety devices.
- Use of single-use disposable equipment

Samples should be marked with a ‘Biohazard/danger of infection’ label to both the container and form and the laboratory informed in advance that a sample is being sent.

2.3 Spillages

Refer to the IPC manual for guidance. It should be noted that none of the methods currently suggested by the World Health Organisation (WHO) for prion inactivation are likely to be fully effective therefore it is essential that standard precautions are adhered to.

2.4 Waste

Tissues and contaminated materials such as dressings and sharps, from patients with, or “at increased risk” of, CJD/vCJD should be disposed of as in the following table:

Table 1: Disposal of clinical waste from patients with, or “at increased risk” of CJD or vCJD

<table>
<thead>
<tr>
<th>Diagnosis of CJD</th>
<th>High or medium risk tissue* (Orange bag)</th>
<th>Low risk tissue and body fluids** (Yellow tiger stripe bag - offensive)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Definite</td>
<td>Alternative Treatment or Incinerate</td>
<td>Normal clinical waste disposal</td>
</tr>
<tr>
<td>Probable</td>
<td>Alternative Treatment or Incinerate</td>
<td>Normal clinical waste disposal</td>
</tr>
<tr>
<td>“At increased risk”</td>
<td>Alternative Treatment or Incinerate</td>
<td>Normal clinical waste disposal</td>
</tr>
</tbody>
</table>

** Tissues and materials deemed to be low risk include body fluids such as urine, saliva, sputum, blood, and faeces. Blood from
vCJD patients is considered to be low risk except when transfused in large volumes.

Sharps should be disposed of into the appropriate sharps bin (yellow or cytotoxic).

2.5 After Death

Following the death of the patient they should be released to the care of the Funeral Director. Standard precautions must continue to be applied during last offices procedures. It is recommended that the deceased is placed in a body bag, which should be labelled as ‘High-Risk or Danger of Infection’ prior to removal by the Funeral Director, in line with standard procedures for deceased patients where there is a known infection risk. The infection control section on the notification of death form (WZT030) must be completed and a copy given to the undertakers concerned. Refer to the Royal Marsden’s Last Offices Procedure and the Trust’s policy for the Transfer of the Deceased into the care of another Service Provider for further guidance.

Post-mortem examination may be required to confirm a clinical diagnosis and cause of death in patients with suspected CJD/vCJD.

2.6 Caring for symptomatic patients at home

Staff caring for patients in their own homes should adhere to standard infection prevention and control precautions. Families caring for patients at home should be advised of the standard infection prevention and control practices. It is recommended that they are provided with disposable gloves, plastic aprons, face protection, paper towels, waste bags and sharps containers, as appropriate. Provision should be made for the removal and disposal of infectious clinical waste and sharps from the home. None infectious waste can, with the homeowners permission, be disposed of in the patients own household waste bin. Refer to the Trust’s Waste Management Policy for further guidance.

It is assumed that all body fluid spillages in the community will be classified as low risk material, for example blood and urine. Refer to the Trust Blood and Body Fluid Spillage policy and ensure standard infection prevention and control precautions are adhered to.

Clinical waste should be disposed of as identified in Table 1 and in accordance with the Trust Waste Management policy.

Patients’ clothes and bed linen can be washed as normal, although in the interests of general hygiene it is recommended that contaminated linen is laundered separately.

2.7 Occupational Exposure

Although cases of CJD/vCJD have been reported in healthcare workers, there have been no confirmed cases linked to occupational exposure.
However, it is prudent to take a precautionary approach.

The highest potential risk in the context of occupational exposure is from exposure to high infectivity tissues through direct inoculation, for example as a result of sharps injuries, puncture wounds or contamination of broken skin, and exposure of the mucous membranes.

Compliance with standard infection IPC precautions will help to minimise risks from occupational exposure.

Refer to the Sharps Policy - Safe Use and Disposal of Sharps and Management of Contamination Injuries for actions to be taken following contamination injuries.

2.8 List of workers exposed to TSE agents

Control of Substances Hazardous to Health (COSHH) regulations require employers to keep a list of employees who work with TSE agents. A local risk assessment should be completed to determine whether a list is required for staff undertaking specific procedures.

3. Definitions/Explanation of Terms Used

Prions - abnormal, pathogenic agents that are transmissible and are able to induce abnormal folding of specific normal cellular proteins called prion proteins that are found most abundantly in the brain. The functions of these normal prion proteins are still not completely understood. The abnormal folding of the prion proteins leads to brain damage and the characteristic signs and symptoms of the disease. Prion diseases are usually rapidly progressive and always fatal.

Sporadic CJD - is the most common type of CJD. The precise cause is unclear, but it has been suggested that in some people a normal brain protein undergoes an abnormal change (misfolding) and turns into a prion.

Variant CJD - is likely to be caused by consuming meat from a cow that has been infected with a similar prion disease called Bovine Spongiform Encephalopathy (BSE) - also known as 'mad cow disease'.

4. RESPONSIBILITIES, ACCOUNTABILITIES AND DUTIES

4.1 Refer to the home page, section 4 of the Infection Prevention and Control policy/manual and overarching Equality Impact Assessment'.

5. LINKS TO ANY ASSOCIATED DOCUMENTS

6. REFERENCES

- COSHH Regulations - Control of Substances Hazardous to Health
- Health and Safety at Work Act (1974)
- Minimise transmission risk of CJD and vCJD in healthcare settings - GOV.UK
- Management of Health and Safety at Work Regulations (1999)

7. APPENDICES

To access the following Appendices please see IPC Manual homepage https://www.rdash.nhs.uk/46192/infection-prevention-and-control-manual/


Appendix 41 - Distribution of TSE infectivity in human tissues and body fluids