Clinical

Creutzfeldt-Jakob Disease (CJD): Standard Operating Procedure

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Contents

1. Introduction 3
2. Purpose 3
3. Scope 3
4. Clinical features 3
5. Patient categorisation 4
6. Care of CJD/vCJD patients 6
7. Categorisation of body tissues 6
8. Sample taking and other invasive medical procedures 7
9. Biopsy samples 8
10. Spillages 9
11. Clinical Waste 9
12. Bed linen 10
13. Occupational Exposure 10
14. After Death 11
15. Standard precautions in the home setting 11
16. Where to get help and advice: 11
17. Process For Monitoring Compliance And Effectiveness 12
18. References 13

Change Control – Amendment History

<table>
<thead>
<tr>
<th>Version</th>
<th>Dates</th>
<th>Amendments</th>
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<tbody>
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Page 2 of 13
1. Introduction

Creutzfeldt-Jakob Disease (CJD) and related disorders, belong to group of diseases known as Transmissible Spongiform Encephalopathies (TSEs) or prion diseases. These are fatal degenerative brain diseases which occur in humans and some animal species. Bovine spongiform encephalopathy (BSE) in cattle and scrapie in sheep also belong to this group of diseases.

The Human CJD Related Disorders are:

- Creutzfeldt-Jacob disease (CJD)
  - classical sporadic
  - familial;
  - iatrogenic and
  - variant CJD (vCJD)
- Gerstmann-Straussler-Scheinker syndrome (GSS)
- Fatal familial insomnia (FFI)
- Kuru
- Variably Protease-Sensitive Prionopathy (VPSPr)

The causative agent is remarkably resistant to conventional sterilisation and disinfection techniques. It is thought that CJD is caused by infectious proteins known as ‘prions’ (Prp-res), which are rogue forms of a normal protein found in the brain.

Whilst the evidence to date does not suggest that CJD or related disorders are spread from person to person by close contact, it is known that transmission can occur in specific situations associated with medical interventions e.g. hormones sourced from human pituitary glands, via blood transfusion of non-leucodepleted red blood cells, through plasma products and neurosurgery with inadequately decontaminated instruments. Consequently, certain procedures need to be followed to identify patients who present a greater than average risk of carrying a CJD or related agent and thereafter managing their care appropriately, whilst reducing the risk of on-going transmission to staff and other patients. There have been no known transmissions of vCJD via surgery or use of tissues or organs.

2. Purpose

Patients and staff have a right to be protected from preventable infections and healthcare staff have a duty to safeguard the well being of their patients. The Health Act 2008 requires all NHS bodies to have in place appropriate core policies of which CJD is one. The purpose of this policy is to provide guidance about the management and control of CJD within the healthcare setting.

3. Scope

This document applies to all South Staffordshire and Shropshire employees and all those visiting the Trusts premises such as contractors, agency/bank/locum staff, students and volunteers.

4. Clinical features

CJD is invariably fatal. The illness usually has a short duration after the onset of progressive symptoms but varies according to the type of CJD. The median illness duration is approximately 3-4 months in classical CJD, 14 months in vCJD and 2-5 years in inherited forms. Clinical feature vary depending on the regions of the brain affected but all patients experience very rapid deterioration.
The common features include:

- Personality change
- Psychiatric symptoms
- Cognitive impairment
- Neurological deficits, including sensory and motor impairments and ataxia
- Myoclonic jerks, or, less frequently, chorea or dystonia
- Rapid, or unpredictable stepwise, deterioration
- Increasing difficulty with communication, mobility, swallowing and continence
- Coma
- Death

Uncertainties about the diagnosis

Because of the rarity of the disease and the lack of a simple diagnostic test it is often difficult to confirm the diagnosis. Older patients with sporadic CJD may initially be given a diagnosis simply of dementia. Younger patients with variant CJD have often initially been given a diagnosis of depression. It should be stressed that many patients may benefit from skilled psychiatric management. However many patients’ families are angry that their relatives have been given a psychiatric diagnosis and managed in a psychiatric setting. This has sometimes proved a barrier in later communication with the family and is another factor making coordination of care more difficult. Professionals should be aware of these issues and acknowledge families’ feelings.

At the moment, a CJD diagnosis can be confirmed only by histological examination of the brain following a brain biopsy, or after a post-mortem. If someone has symptoms suggestive of variant CJD (vCJD), a full neurological examination would be conducted by a specialist. There is no proven treatment or cure for CJD, and the disease leads to death. Research is being carried out on the causes, tests and possible treatments for the disease.

The National CJD Research and Surveillance Unit carries out surveillance of CJD throughout the UK and provides further information on CJD for clinicians and members of the public on its website. This includes information on diagnostic criteria, the number of cases, epidemiology, research and the latest short-term incidence projections.

5. Patient categorisation

When considering measures to prevent transmission to patients or staff in the healthcare setting, 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 (see Annex B for full diagnostic criteria), 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.

In most routine clinical contact, no additional precautions are needed for the care of patients in the “increased risk” patient groups. However, when certain invasive interventions are performed, there is the potential for exposure to the agents of TSEs. In these situations it is essential that control measures are in place to prevent iatrogenic CJD/vCJD transmission.

Categorisation of patients by risk

<table>
<thead>
<tr>
<th>Patient groups</th>
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<tbody>
<tr>
<td>Symptomatic patients</td>
<td>• Patients who fulfill the diagnostic criteria for definite, probable or possible CJD or vCJD (see Annex B for diagnostic criteria)</td>
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</table>
• Patients 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

**Patients “at increased risk” from genetic forms of CJD**

• Individuals who have been shown by specific genetic testing to be at significant risk of developing CJD.
  
  • Individuals who have a blood relative known to have a genetic mutation indicative of genetic CJD;
  
  • Individuals who have or have had two or more blood relatives affected by CJD or other prion disease

**Patients identified as “at increased risk” of CJD/vCJD through iatrogenic exposures**

• Recipients of hormone derived from human pituitary glands, e.g. growth hormone, gonadotrophin. In the UK the use of human-derived gonadotrophin was discontinued in 1973, and use of cadaver-derived human growth hormone was banned in 1985. However, use of human-derived products may have continued in other countries after these dates.
  
  • Individuals who underwent intradural neurosurgical or spinal procedures before August 1992. These patients may have received a graft of human-derived dura mater and should be treated as being “at increased risk” unless evidence can be provided that human-derived dura mater was not used. Patients who received a graft of human-derived dura mater before August 1992 are “at increased risk” of transmission of sporadic CJD.
  
  • Individuals who have had surgery using instruments that had been used on someone who went on to develop CJD/vCJD, or was “at increased risk” of CJD/vCJD;
  
  • Individuals who have received an organ or tissue from a donor infected with CJD/vCJD or “at increased risk” of CJD/vCJD;
  
  • Individuals who have been identified prior to high risk surgery as having received blood or blood components from 80 or more donors since January
Recipients of ocular transplants, including corneal transplants, are not considered to be “at increased risk” of CJD/vCJD.

All people who are “at increased risk” of CJD/vCJD are asked to help prevent any further possible transmission to other patients by following this advice:

- Don’t donate blood. No-one who is “at increased risk” of CJD/vCJD, or who has received blood donated in the United Kingdom since 1980, should donate blood;
- Don’t donate organs or tissues, including bone marrow, sperm, eggs or breast milk;
- If you are going to have any medical, dental or surgical procedures, tell whoever is treating you beforehand so they can make special arrangements for the instruments used to treat you if you need certain types of surgery or investigation;
- You are advised to tell your family about your increased risk. Your family can tell the people who are treating you about your increased risk of CJD/vCJD if you need medical or surgical procedures in the future and you are unable to tell them yourself.

6. Care of CJD/vCJD patients

Available epidemiological evidence does not suggest that normal social or routine clinical contact with a CJD or vCJD patient presents a risk to healthcare workers, relatives and others in the community.

- Isolation of patients with CJD or vCJD is not necessary, and they can be nursed in an open ward using standard infection control precautions in line with those used for all other patients.

7. Categorisation of body tissues

At present, there is no evidence of infectivity in saliva, body secretions or excreta.

Although prion protein has not been detected in the CSF in either sporadic or variant CJD experimental transmission of infectivity has been achieved from CSF in sporadic CJD indicating that levels of infectivity are likely to be much lower than in the central nervous system (CNS).

Blood is considered to be low risk tissue.

In the main, most infectivity is likely to be concentrated in the central nervous tissue. In vCJD, infectivity is also likely to be present in lymphoid tissue, albeit at a lower level.
Risk status of different tissues

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<tr>
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<th>CJD other than VCJD</th>
<th>vCJD</th>
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<tr>
<td><strong>High</strong></td>
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<tr>
<td>Brain</td>
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<tr>
<td>Spinal cord</td>
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<td></td>
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<tr>
<td>Posterior eye</td>
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<td></td>
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<tr>
<td><strong>Medium</strong></td>
<td></td>
<td></td>
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<tr>
<td>Olfactory epithelium*</td>
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<tr>
<td><strong>Low</strong></td>
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<tr>
<td>Anterior eye</td>
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<td>Anterior eye</td>
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</tbody>
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**All other tissues including blood and dental tissues**

**lymphoid tissue refers to the spleen, thymus, tonsils and adenoids, lymph nodes, the appendix and the gastro-intestinal tract sub-mucosa**

8. 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.


It is important to ensure that only 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, body 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 treated as potentially infectious for blood-borne viruses and handled with standard infection control precautions as for any other patient, i.e.;

- use of disposable gloves and eye protection where splashing may occur;
- avoidance of sharps injuries and other forms of parenteral exposure;
- safe disposal of sharps and contaminated waste in line with locally approved arrangements; and
- single-use disposable equipment should be used wherever practicable.
When taking biopsy specimens of medium or high risk tissue, for example tonsil biopsy in a patient with suspected vCJD, every effort should be taken to minimise the risk of infecting the operator or contaminating the environment.

Samples from patients with, or “at increased risk” of, CJD/vCJD should be marked with a ‘Biohazard’ label, and it is advisable to inform the laboratory in advance that a sample is being sent.

**Non-Invasive Procedures**

For non-invasive procedures (e.g. X-Ray or other imaging procedures), no specific precautions other than those that would normally be applied to all patients is required.

**Invasive medical procedures**

An invasive procedure is defined as one where there is a risk of exposure to body tissues.

**General precautions relating to all invasive procedures**

- The unusual resistance of the agents thought to be responsible for CJD and related disorders, means that single-use disposable equipment should be used wherever practicable and all other small items of equipment contaminated whilst undertaking invasive procedures and obtaining specimens should be destroyed.
- Blood, biopsy and lumbar puncture samples from known or suspect patients should only be taken by trained personnel who are aware of the hazards involved.
- Disposable gloves and eye protection (where splashing may occur) should be worn.
- Where procedures are performed at the bedside e.g. a lumbar puncture, care should be taken to ensure the environment can be readily cleaned should a spillage occur.

**The following precautions should be taken for procedures on known or suspect patients if clinical intervention involves High or medium risk tissues:**

- The intervention should be performed in an operating theatre.
- Where possible, procedures should be performed at the end of the list, to allow normal cleaning of theatre surfaces before the next session.
- Only the minimum number of healthcare personnel required should be involved.
- The following protective clothing should be worn by healthcare personnel.
  - Liquid repellent operating gown.
  - Gloves.
  - Mask and goggles, or full face visor.
  - One way flow of instruments should be maintained.
  - Protective clothing should be disposed of as clinical waste.

- Single-use disposable surgical instruments and equipment should be used where possible, and subsequently destroyed by incineration or sent to the instrument store
- Effective tracking of re-usable instruments should be in place, so that instruments can be related to use on a particular patient. Refer to national guidance

**9. Biopsy samples**

The collection of biopsy samples should involve the same precautions used for all work of this type with any patient although particular care should be taken with lymphoid tissue specimens. Disposable gloves, aprons and eye protection (where splashing may occur) should be worn. Single-use disposable instruments should be used where possible, where items that are normally re-usable need to be used.
10. Spillages

The infectious agents associated with CJD and related disorders, are unusually resistant to inactivation techniques, the main defence is efficient removal of the contaminating material and thorough cleaning of the surface.

Surface decontamination and the management of spillages:

- Disposable gloves and an apron should be worn when removing any spillage and these should be disposed of as clinical waste.
- For minor spillages of low risk materials such as blood and urine from definite, probable or high-risk cases, the surface should be disinfected using standard infection control precautions.
- For spillages of larger volumes of low risk liquids, absorbent material should be used to take up the spillage e.g. paper towels. Gently pour the appropriate sodium hypochlorite solution over the towels. Wherever practicable, a contact time of 1hr should be maintained before removing the spillage into a yellow bag for incineration.
- For decontamination of surfaces in contact with high or medium risk material from definite, probable or high risk cases. Sodium hypochlorite containing 10,000ppm available chlorine or 1M sodium hydroxide should be used.
- Care should be taken when handling these chemicals at these concentrations. Simple precautions such as the use of gloves, eye protection and plastic aprons should be taken.
- Needs may differ according to different circumstances but in any event a full risk assessment will be required.
- Repeated wetting with the disinfectant over the 1-hour treatment period is necessary. As this concentration of hypochlorite can be corrosive for some commonly used surface finishes it should be used with caution.
- Any waste (including cleaning tools such as mop heads and PPE worn) should be disposed of as clinical waste.

All materials used in the cleaning operation should be disposed of by incineration. All waste should be disposed of as clinical waste.

11. Clinical 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:

- High or medium risk tissue should be disposed of as Clinical waste in orange bags and by incineration in line with locally approved agreements.
- Low risk tissue and body fluids should be disposed of as clinical waste in yellow and black bags

<table>
<thead>
<tr>
<th>Diagnosis of CJD</th>
<th>High or medium risk tissue*</th>
<th>Low risk tissue and body fluids**</th>
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<tr>
<td>Definite</td>
<td>Incinerate</td>
<td>Normal clinical waste disposal</td>
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<tr>
<td>Probable</td>
<td>Incinerate</td>
<td>Normal clinical waste disposal</td>
</tr>
<tr>
<td>“At increased risk”</td>
<td>Incinerate</td>
<td>Normal clinical waste disposal</td>
</tr>
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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.
12. Bed linen

Used or fouled bed linen (contaminated with body fluids or excreta) should be washed and dried in accordance with current standard practice and advice for infected linen. No further handling or processing requirements are necessary.

13. 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 (e.g. as a result of “sharps” injuries, puncture wounds or contamination of broken skin), and exposure of the mucous membranes (e.g. conjunctiva) should also be avoided.

Compliance with standard infection control precautions, in line with the trust SOP will help to minimise risks from occupational exposure.

Healthcare personnel, who work with patients with definite, probable or possible CJD or vCJD, or with potentially infected tissues, should be appropriately informed about the nature of the risk and relevant safety procedures.

For any accident involving “sharps”, or contamination of abrasions with blood or body fluid(s), wounds should be gently encouraged to bleed, gently washed (avoid scrubbing) with warm soapy water, rinsed, dried and covered with a waterproof dressing, or further treatment given appropriate to the type of injury. Splashes into the eyes or mouth should be dealt with by thorough irrigation. The accident should be appropriately reported and an adverse incident form completed.

List of workers exposed to TSE agents

Under certain circumstances COSHH requires employers to keep a list of employees who are exposed to HG3 or 4 agents. The decision to keep a list depends on the local risk assessment. This should be carried out by senior manager responsible for the area concerned with advice from the occupational health Physician or Infection Control Doctor.

For TSE agents a list is only required where employees deliberately work with the agent. For example:

Those involved in laboratory research work and veterinary clinical work with a TSE agent.

Staff performing invasive clinical procedures on patients suspected to be suffering from CJD of any type, particularly where there is a risk of exposure to central nervous tissue, eye tissue or other tissues known to contain CJD infectivity.

Healthcare personnel who work with patients with definite, probable or possible CJD/vCJD, or with potentially infected tissues, should be appropriately informed about the nature of the risk and relevant safety procedures.

The routine clinical care of patients with CJD or a related disorder is unlikely to pose a significant risk of exposure to CJD of any type and staff working with such patients would not need to be included on such a list.

In cases of unintentional exposure, a list may be required if the risk assessment shows that there is a significant risk.

The risk is deemed to be significant if more than basic hygiene measures are necessary to protect staff or if the control measures listed in COSHH are specifically applied. The list should be kept where there is a likelihood of exposure and not simply when there has been a known incident or accident, although it should also include details of these.

Recording details of incidents or accidents on this list is not the same as the requirement to report certain diseases and accidents to HSE under the Reporting of Injuries, Diseases and Dangerous Occurrences Regulations (RIDDOR).
The information that should be recorded includes:
The type of work done and where known, any specific exposure, accident or incident. Because of the long latency period of TSE agents and their serious long-term sequelae, the list must be kept for 40 years after the last known exposure.

This list is in addition to the health record (which is required for the purposes of health surveillance under COSHH or MHSWR) and must be made available to any doctor appointed to carry out health surveillance, e.g. the occupational health physician. It must also be available to any employee who is specifically responsible for health and safety.

14. After Death

Normal infection control procedures apply. The deceased patient must be placed in a body bag with ‘Danger of Infection’ label applied and moved to the mortuary using standard precautions.

Viewing the deceased
Relatives of the deceased may wish to view or have some final contact with the body. Such viewing and possible superficial contact, such as touching or kissing, need not be discouraged even if a post-mortem has taken place. Body bags may be rolled down temporarily to allow superficial contact; there is no need to deny the relatives this opportunity if a post-mortem has been performed.
Following post mortem examination it is advisable to minimise contact, particularly in circumstances where penetrating injuries could arise.

15. Standard precautions in the home setting

Although CJD is not thought to present risk through normal social or routine clinical contact, families caring for patients at home should be advised of the standard infection control practice that would apply to any patient. They should be provided with gloves, paper towels, bags and sharps containers as appropriate.
Families should not be dissuaded from ordinary contact with patients but should wear gloves and aprons if handling body fluids.
In the home setting, patients’ clothes and bed linen may be washed as normal, but fouled linen should not be washed with other laundry. Families should be provided with plastic bags for any clinical waste materials and sharps containers if appropriate. Provision should be made for the removal of clinical waste and sharps from the home for incineration.
Spillages of body fluids, including blood, should be removed using absorbent towels (e.g. kitchen paper) and the surface washed thoroughly with detergent and warm water. Disposable gloves and apron should be worn. Health professionals involved in caring for the patient should be in contact with the local authority to ensure that appropriate arrangements for the removal and disposal of waste are put in place.

16. Where to get help and advice:
Creutzfeldt-Jakob Disease: Guidance for Healthcare Workers” advice on the care of patients with CJD/vCJD is available at


National CJD Surveillance Unit   http://www.cjd.ed.ac.uk/
17. Process For Monitoring Compliance And Effectiveness

This SOP will be reviewed three yearly or earlier in light of new national guidance or other significant change in circumstances.

Compliance with SOP committee/group. The results of the annual audit will be escalated to the appropriate committee/group where appropriate.
<table>
<thead>
<tr>
<th>Aspect of compliance or effectiveness being monitored</th>
<th>Monitoring method</th>
<th>Individual or department responsible for the monitoring</th>
<th>Frequency of the monitoring activity</th>
<th>Group/committee/ forum which will receive the findings/monitoring report</th>
<th>Committee/individual responsible for ensuring that the actions are completed</th>
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<tbody>
<tr>
<td>Compliance with Infection Prevention and control policies and practices</td>
<td>Annual Infection Prevention and control audits</td>
<td>Audit Department</td>
<td>Yearly</td>
<td>Infection Control Committee</td>
<td>Matrons and Ward Managers</td>
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<tr>
<td>Organisation’s expectations in relation to staff training, as identified in the training needs analysis</td>
<td>Training Reports</td>
<td>Learning and Development Department</td>
<td>Monthly</td>
<td>HRODE Committee</td>
<td>HRODE Committee</td>
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18. References

Creutzfeldt-Jakob Disease: Guidance for Healthcare Workers” advice on the care of patients with CJD/vCJD is available at