journey to change

NHS
Tees, Esk and Wear Valleys
NHS Foundation Trust

Public – To be published on the Trust external website

CJD (Creutzfeldt-Jakob Disease) and patient management

Ref: IPC-0001-003-v5

Status: Approved
Document type: Procedure.
Overarching policy: Infection Prevention Control Policy

Contents

1 Introduction

This procedure is required to inform staff in the management of caring for patients with suspected or confirmed CJD.

This procedure supports Our Journey To Change (OJTC) as set out in the overarching **Infection Prevention Control Policy.**

2 Purpose

Following this procedure will help the Trust to:-

- Provide a safe environment for both staff and patients and to manage cases of CJD safely and effectively.
- Ensure that any patient suspected on clinical grounds of having any type of CJD, following discussion with a local neurologist, is reported to the National Creutzfeldt Jakob Disease Research & Surveillance Unit (NCJDRSU).

3 Who this procedure applies to

This procedure applies to all healthcare professionals within Tees, Esk and Wear Valleys NHS Foundation Trust, in the care and management of patients who are known or suspected to have CJD.

4 Related documents

This procedure describes what you need to do to implement the CJD section of the Infection Prevention and Control Policy .

The Standard Precautions for Infection Prevention and Control defines the universal standards for IPC which you **must** read, understand and be trained in before carrying out the procedures described in this document.

This procedure also refers to:-

- ✓ Accidental Inoculation
- ✓ Decontamination of Equipment
- ✓ Waste management policy

5 How does CJD affect people?

Creutzfeldt-Jakob disease (CJD) is one of a rare group of diseases, known as transmissible spongiform encephalopathies (TSEs), which affect the structure of the brain. TSEs cause dementia and a range of neurological symptoms, including ataxia, myoclonus, chorea or dystonia.

Creutzfeldt-Jakob disease (CJD) is caused by an abnormal infectious protein in the brain called a prion.

TSEs are recognised in both animals and humans. In animals, the best-known TSE is bovine spongiform encephalopathy (BSE or “mad cow disease”).

In humans, there are four main types of CJD:

- Sporadic CJD
- Variant CJD
- Genetic CJD and other inherited prion diseases
- Iatrogenic CJD.

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.

5.1 Incubation period

- The illness usually has a short duration after the onset of progressive symptoms but varies according to the type of CJD.
- Clinical features vary depending on the regions of the brain affected but all patients experience very rapid deterioration.
- CJD is always fatal.

5.2 Signs and symptoms

- Personality change.
- Psychiatric symptoms.
- Cognitive impairment.
- Neurological deficits including sensory and motor impairments and ataxia.
- Myoclonic jerks or, less frequently, chorea and dystonia.
- Rapid or unpredictable stepwise deterioration.

- Increasing difficulty with communication, mobility, swallowing and continence.
- Coma.
- Death.

5.3 Person-to-person spread.

- Normal social or routine clinical contact with patients suffering from any type of CJD **does not** present a risk to health care workers, relatives, or the community.
- Because prions are resistant to conventional sterilisation/disinfection techniques, there is a theoretical risk of transmission during certain surgical procedures.

5.4 Occupational exposure

Although there have been no confirmed cases of CJD/vCJD linked to occupational exposure, you **must** still be cautious when dealing with patients with suspected or confirmed CJD/vCJD.

The highest potential risk is from exposure to high infectivity tissues through direct inoculation e.g. because of 'sharps' injuries, puncture wounds or contamination of broken skin), and exposure of mucous membranes.

Comply with standard infection control precautions to minimise risks from occupational exposure.

Treat and report inoculation accidents/injuries as defined in the Accidental Inoculation document.

5.5 Risks of contracting CJD?

UKHSA (2018) advise that eating beef or beef products from BSE infected cattle is the most likely cause of vCJD, and most of the people in the UK who have CJD would have been exposed in this way. Other potential sources of CJD infection include contaminated medical equipment or infected transplant material.

Prion diseases like CJD can spread from one person to another only in certain circumstances through healthcare. They are not infectious in usual ways, e.g. by coughing or sneezing, touching or by having sex, nor is there evidence that the disease can spread during pregnancy to the unborn baby or through breastfeeding.

5.5.1 Sporadic CJD

Even though sporadic CJD is very rare, it's the most common type of CJD, accounting for around 8 in every 10 cases. It's not known what triggers sporadic CJD, but it may be that a normal prion protein spontaneously changes into a prion, or a normal gene spontaneously changes into a faulty gene that produces prions. Sporadic CJD is more likely to occur in people who have specific versions of the prion protein gene. At present, nothing else has been identified that increases the risk of developing sporadic CJD.

5.5.2 Variant CJD

There's clear evidence that variant CJD (vCJD) is caused by the same strain of prions that causes bovine spongiform encephalopathy (BSE, or "mad cow" disease). In 2000, a government inquiry concluded that the prion was spread through cattle that were fed meat-and-bone mix containing traces of infected brains or spinal cords. The prion then ended up in processed meat products, such as beef burgers, and entered the human food chain. Strict controls have been in place since 1996 to prevent BSE entering the human food chain, and the use of meat-and-bone mix has been made illegal. It appears not everyone who's exposed to BSE-infected meat will go on to develop vCJD.

5.5.3 Familial or inherited CJD

Familial or inherited CJD is a rare form of CJD caused by an inherited mutation (abnormality) in the gene that produces the prion protein. The altered gene seems to produce misfolded prions that cause CJD. Everyone has 2 copies of the prion protein gene, but the mutated gene is dominant.

This means you only need to inherit 1 mutated gene to develop the condition. So if 1 parent has the mutated gene, there's a 50% chance it will be passed on to their children. As the symptoms of familial CJD don't usually begin until a person is in their 50s, many people with the condition are unaware that their children are also at risk of inheriting this condition when they decide to start a family.

5.5.4 Iatrogenic CJD

Iatrogenic CJD (iCJD) is where the infection is spread from someone with CJD through medical or surgical treatment.

6 Dealing with new or suspected cases of CJD

6.1 Who to notify.

Event/situation	Action to be taken	Who by
On suspicion of a patient having any type of CJD	Discuss with a local consultant neurologist	Clinician in charge of the patient
	Inform the National CJD Research & Surveillance Unit (NCJDRSU) 0131 537 1980 (Edinburgh) and local health protection team.	Consultant neurologist

	Visit and examine the patient	NCJDRSU neurologist
	Complete a detailed risk factor questionnaire with a relative	NCJDRSU research nurse

7 Infection Prevention and Control precautions for a patient known or suspected to have CJD.

- Normal social or routine clinical contact does not present a risk to healthcare workers, families, or others.
- Isolating patients with CJD or vCJD is not necessary. Patients with CJD may be nursed on an open ward and continue with normal activities unless other condition requires isolation.
- No special precautions are needed other than standard infection control practice that would apply to any other patient. This includes the need for handwashing before and after any procedure and cleaning of all multiuse medical equipment with detergent and water/ detergent wipes following each use.
- The use of gloves and aprons when blood and body fluids are involved.
- Linen should be handled as standard. Only linen that becomes contaminated with blood, CSF or tissue fluids should be treated as 'infected linen' in accordance with laundry guidance. If such contamination is likely, consider disposable linen.
- Clinical waste requires incineration and should be disposed of wearing gloves and aprons into a yellow rigid bin.
- Single use equipment should be used for wound dressings and clinical waste treated as above.
- Spillages of blood and body fluids should be dealt with immediately using a chlorine releasing agent of 10,000 ppm gloves and aprons must be worn when handling spillages. Equipment used (including PPE and mops / cloths) must be disposed of as clinical waste for incineration.

7.1 Taking specimens

- Only trained personnel who know the hazards involved must take blood and biopsy specimens. The receiving pathology laboratory must be informed prior to any samples being taken.
- When collecting blood specimens, take the same precautions as for all work of this type with **any** patient i.e.:
  - avoiding sharps injuries and other forms of parenteral exposure.
  - disposing of sharps and contaminated waste in line with Trust policy.

- Label specimens with a 'Danger of Infection' label, and it is advisable to inform the laboratory in advance that a specimen is being sent.

7.2 Dealing with equipment

- Clean re-usable clinical care equipment such as commodes, wheelchairs, hoists etc. using normal procedures. (See Decontamination of Equipment).
- Dispose of single-use items into clinical waste for incineration.

7.3 Surgical invasive procedures

Notify the receiving trust/organization where the surgery or invasive procedures are planned to take place.

!	When any surgery including endoscopy is anticipated the Infection Prevention and Control Team **must** be contacted.
!	When 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.
!	The patient's clinician and the infection prevention and control team will liaise with the relevant acute NHS Trust to ensure that patients with, suspected or at risk of CJD undergo a pre-surgery assessment.

7.4 Dentistry

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 patient with, or 'at increased risk' of CJD or vCJD, should be refused routine dental equipment. Such patients can be treated in the same way as any other patient.

7.5 Death

- After death inform the infection prevention control team, ward procedure for last offices is the same as for any other patient.
- Relatives need not be discouraged from viewing the body or from superficial contact such as touching the face.
- Place the body in a body (cadaver) bag and apply 'Danger of infection' stickers to the wrist band, shroud, and mortuary card
- Inform the undertakers of known or suspected diagnosis of CJD.

8 Definitions

Term	Definition
Classical CJD	Thought to affect the central nervous system and the brain, spinal cord and eyes are thought to be potentially infectious.
Dura mater	The tough outermost membrane of the three that cover the brain and spinal cord.
NCJDRSU	National Creutzfeldt Jakob Disease Research & Surveillance Unit
Prions	The precise cause of CJD is unknown but infectious proteins known as 'prions' is a likely cause. Normal prion proteins are found in the tissues of healthy people. Those causing disease to alter in shape by folding in an abnormal way. This abnormally shaped protein then influences the normal prion proteins to alter their shape. The accumulation of this altered prion protein causes destruction of nervous tissue and the clinical manifestations of the disease.
Variant CJD (vCJD)	As well as affecting the central nervous system, vCJD has also been detected in lymphatic tissue such as tonsil and appendix. vCJD poses a greater potential risk of person-to-person spread in healthcare settings than classical CJD.
TSEs	CJD refers to Transmissible Spongiform Encephalopathies (TSEs), which are characterised by degeneration of the nervous system and degenerative brain disease, which are invariably fatal.

9 How this procedure will be implemented

- This procedure will be published on the Trust's intranet and external website.
- Line managers will disseminate this procedure to all Trust employees through a line management briefing.

9.1 Training needs analysis

Staff/Professional Group	Type of Training	Duration	Frequency of Training

Clinical staff	Face to face	1 hour	Available on request, provided by IPC team

10 How the implementation of this procedure will be monitored

	Auditable Standard/Key Performance Indicators	Frequency/Method/Person Responsible	Where results and any Associate Action Plan will be reported to, implemented and monitored; (this will usually be via the relevant Governance Group).
1	Any identified patient will be reported to IPC by the clinical team	As soon as diagnosis is known/suspected	IPCC

11 References

Department of Health(2012) Minimise transmission risk of CJD and vCJD in healthcare setting: Prevention of CJD and vCJD by Advisory Committee on Dangerous Pathogens' Transmissible Spongiform Encephalopathy (ACDP TSE) Subgroup. Transmissible Spongiform Encephalopathy Agents: Safe Working and the Prevention of Infection: Part 4

https://assets.publishing.service.gov.uk/government/uploads/system/uploads/attachment_data/file/427854/Infection_controlv3.0.pdf

Transmissible Spongiform Encephalopathy Agents: Safe Working and the Prevention of Infection. Guidance from the Advisory Committee on Dangerous Pathogens and the Spongiform Encephalopathy Advisory Committee. 1998, 2003, 2004 and 2006.

National Institute for Health and Clinical Excellence. (2020) Reducing the risk of transmission of Creutzfeldt–Jakob disease (CJD) from surgical instruments used for interventional procedures on high-risk tissues. Overview | Reducing the risk of transmission of Creutzfeldt–Jakob disease (CJD) from surgical instruments used for interventional procedures on high-risk tissues | Guidance | NICE

CJD: information leaflets for patients and healthcare professionals - GOV.UK (www.gov.uk)

Transmissible spongiform encephalopathy agents: safe working and the prevention of infection Frequently asked questions (publishing.service.gov.uk)

The National CJD Research & Surveillance Unit https://www.cjd.ed.ac.uk/

Public Health England (2018) Patients at increased risk of Creutzfeldt-Jakob disease (CJD): background Information for healthcare staff (publishing.service.gov.uk)

12 Document control (external)

To be recorded on the policy register by Policy Coordinator

Required information type	Information
Date of approval	13 February 2025
Next review date	13 February 2028
This document replaces	CJD (Creutzfeldt-Jakob Disease) and patient management Ref IPC-0001-003-v4.1
This document was approved by	IPCC
This document was approved	13 February 2025
This document was ratified by	n/a
This document was ratified	n/a
An equality analysis was completed on this policy on	03 January 2025
Document type	Public
FOI Clause (Private documents only)	n/a

Change record

Version	Date	Amendment details	Status
4	19 Jan 2022	Full review with minor changes. Transferred to new template. Hyperlinks updated. OJTC text added. References updated	Withdrawn
5	13 Feb 2025	Full review with minor changes. Hyperlinks updated. Additional information added to section 5.5. Phone number added for National CJD Research & Surveillance Unit.	Approved

Appendix 1 - Equality Impact Assessment Screening Form

Please note: The Equality Impact Assessment Policy and Equality Impact Assessment Guidance can be found on the policy pages of the intranet

Section 1	Scope
Name of service area/directorate/department	Infection prevention and Control
Title	CJD (Creutzfeldt-Jakob Disease) and patient management
Type	Procedure/guidance*
Geographical area covered	Trust wide
Aims and objectives	To set standards in practice to ensure the delivery of patient care is carried out safely and effectively by the trust staff. To comply with the HCAI Code of Practice of the Health and Social Care Act 2008.
Start date of Equality Analysis Screening	13 December 2024
End date of Equality Analysis Screening	03 January 2025

Section 2	Impacts
Who does the Policy, Procedure, Service, Function, Strategy, Code of practice, Guidance, Project or Business plan benefit?	Staff, patients and relatives.
Will the Policy, Procedure, Service, Function, Strategy, Code of practice, Guidance, Project or Business plan impact negatively on any of the protected characteristic groups? Are there any Human Rights implications?	• **Race** (including Gypsy and Traveller) **NO** • **Disability** (includes physical, learning, mental health, sensory and medical disabilities) **NO** • **Sex** (Men and women) **NO** • **Gender reassignment** (Transgender and gender identity) **NO** • **Sexual Orientation** (Lesbian, Gay, Bisexual, Heterosexual, Pansexual and Asexual etc.) **NO** • **Age** (includes, young people, older people – people of all ages) **NO** • **Religion or Belief** (includes faith groups, atheism and philosophical beliefs) **NO** • **Pregnancy and Maternity** (includes pregnancy, women / people who are breastfeeding, women / people accessing perinatal services, women / people on maternity leave) **NO** • **Marriage and Civil Partnership** (includes opposite and same sex couples who are married or civil partners) **NO** • **Armed Forces** (includes serving armed forces personnel, reservists, veterans and their families) **NO** • **Human Rights Implications NO** (Human Rights - easy read)
Describe any negative impacts / Human Rights Implications	N/A
Describe any positive impacts / Human Rights Implications	N/A

Section 3	Research and involvement
What sources of information have you considered? (e.g. legislation, codes of practice, best practice, nice guidelines, CQC reports or feedback etc.)	NICE guidelines. UKHSA guidance. Transmissible Spongiform Encephalopathy Agents: Safe Working and the Prevention of Infection
Have you engaged or consulted with service users, carers, staff and other stakeholders including people from the protected groups?	No
If you answered Yes above, describe the engagement and involvement that has taken place	
If you answered No above, describe future plans that you may have to engage and involve people from different groups	N/A

Section 4	Training needs
As part of this equality impact assessment have any training needs/service needs been identified?	No
Describe any training needs for Trust staff	n/a
Describe any training needs for patients	n/a
Describe any training needs for contractors or other outside agencies	n/a

Check the information you have provided and ensure additional evidence can be provided if asked.

Appendix 2 – Approval checklist

To be completed by lead and attached to any document which guides practice when submitted to the appropriate committee/group for consideration and approval.

	Title of document being reviewed:	Yes/No/ Not applicable	Comments
1.	**Title**		
	Is the title clear and unambiguous?	Yes	
	Is it clear whether the document is a guideline, policy, protocol or standard?	Yes	
2.	**Rationale**		
	Are reasons for development of the document stated?	Yes	
3.	**Development Process**		
	Are people involved in the development identified?	Yes	
	Has relevant expertise has been sought/used?	Yes	
	Is there evidence of consultation with stakeholders and users?	Yes	
	Have any related documents or documents that are impacted by this change been identified and updated?	Yes	
4.	**Content**		
	Is the objective of the document clear?	Yes	
	Is the target population clear and unambiguous?	Yes	
	Are the intended outcomes described?	Yes	
	Are the statements clear and unambiguous?	Yes	
5.	**Evidence Base**		
	Is the type of evidence to support the document identified explicitly?	Yes	
	Are key references cited?	Yes	
	Are supporting documents referenced?	Yes	
6.	**Training**		
	Have training needs been considered?	Yes	

	Title of document being reviewed:	Yes/No/ Not applicable	Comments
	Are training needs included in the document?	Yes	
7.	**Implementation and monitoring**		
	Does the document identify how it will be implemented and monitored?	Yes	
8.	**Equality analysis**		
	Has an equality analysis been completed for the document?	Yes	
	Have Equality and Diversity reviewed and approved the equality analysis?	Yes	03 Jan 2025 ah
9.	**Approval**		
	Does the document identify which committee/group will approve it?	Yes	
10.	**Publication**		
	Has the document been reviewed for harm?	Yes	
	Does the document identify whether it is private or public?	Yes	Public
	If private, does the document identify which clause of the Freedom of Information Act 2000 applies?	N/A	
11.	**Accessibility** (See intranet accessibility page for more information)		
	Have you run the Microsoft Word Accessibility Checker? (Under the review tab, 'check accessibility'. You must remove all errors)	Yes	
	Do all pictures and tables have meaningful alternative text?	Yes	
	Do all hyperlinks have a meaningful description? (do not use something generic like 'click here')	Yes	