journey to change

NHS
Tees, Esk and Wear Valleys
NHS Foundation Trust

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Title: CJD (Creutzfeldt-Jakob Disease) and patient management

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1 Introduction

In this section describe:

- This procedure is required to inform staff in the management of caring for patients with suspected or confirmed CJD
- This procedure links to Our Journey To Change (OJTC) and has been developed with OJTC in mind, by following the latest guidelines and evidence based practice our aim is to provide a safe environment for both staff and patients and to manage cases of CJD safely and effectively.

To co-create a great experience for our patients, carers and families, so you will experience:

- **Outstanding** and compassionate care, all of the time.
- **Access** to the care that is right for you.
- **Support** to achieve your goals.
- **Choice** and control.

To co-create a great experience for our colleagues, so you will be:

- **Proud**, because your work is meaningful.
- **Involved** in decisions that affect you.
- **Well led** and managed.
- That your workplace is **fit for purpose**.

To be a great partner, so we will:

- Have a **shared understanding** of the needs and the strengths of our communities
- Be **working innovatively** across organisational boundaries to improve services.
- Be **widely recognised** for what we have achieved together.

2 Purpose

Following this procedure will help the Trust to:-

- 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.

Respect

- Listening

Compassion

- Kind

Responsibility

- Honest

- Inclusive
- Working in partnership
- Supportive
- Recognising and Celebrating
- Learning
- Ambitious

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.

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. as a result 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 Who is at risk of contracting CJD?

Public Health England (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, eg 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.

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 NCJDRSU and local health protection unit	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 20,000 ppm gloves and aprons must be worn when handling spillages and equipment used 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.
- 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.
- Safe labelling of specimens
  - ✓ Ensure each specimen is clearly labelled with the patient's name, date of birth, NHS number and location eg. ward name.

- ✓ The pathology request form must also identify the patients details as well as relevant clinical details, reason for the specimen request and any current antibiotic treatment.
- ✓ Ensure the laboratory request form is also signed by the clinician who has requested the specimen.
- ✓ The specimen must be secured in the specimen container and placed into a leak proof sealed specimen bag along with the request form.
- ✓ Any specimens deemed as high risk of infection (e.g. from patients with blood borne viruses or diseases such as Creutzfeldt-Jacob Disease) must be placed into a mini grip plastic bag before being placed into the bag with the pathology request form, they should also be labelled as 'high risk' (high risk stickers can be ordered via cardea).
- ✓ Unlabelled or incorrectly labelled specimens will be discarded by the receiving laboratory department.

Transportation of laboratory specimens

- ✓ All pathology specimens must be transported in a leak proof, washable container. The container must be secure and must comply with UN 3373 standards.
- ✓ Specimen transport containers must not be left unattended in a patient access area.
- ✓ Specimen transport containers must be cleaned at least weekly, or immediately if they become contaminated.
- ✓ Where specimens are transported to the laboratory by vehicle, the transport specimen container must be placed into a cardboard transport box labelled with both the destination and senders name and address.
- ✓ Each specimen container must be in a separate plastic bag with sufficient material to fully absorb any leakage of the specimen
- ✓ Vehicles used for specimen transportation must be equipped with personal protective equipment and a spill kit. Any spillages must be cleaned immediately, and the specimen requester informed as a further specimen will need to be obtained.

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' are a likely cause. Normal prion proteins are found in the tissues of healthy people. Those causing disease 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. (2006) Patient safety and reduction of risk of transmission of Creutzfeldt-Jakob disease (CJD) via interventional procedures. Patient safety and reduction of risk of transmission of Creutzfeldt–Jakob disease (CJD) via interventional procedures | 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

Date of approval:	24 January 2022 v4 19 January 2023 v4.1	
Next review date:	24 January 2025	
This document replaces:	CJD (Creutzfeldt-Jakob Disease) and patient management Ref IPC-0001-003-v3	
This document was approved by:	Name of committee/group	Date
	IPCC (virtual approval)v4 IPCC (amendment approved)v4.1	24 January 2022 19 January 2023
This document was ratified by:	Name of committee/group	Date
	IPCC	Actual amended document to be retrospectively approved at IPCC 20 April 2023
An equality analysis was completed on this document on:	21 January 2022	
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	Approved.
4.1	19 Jan 2023	Minor change only to reflect Information regarding safe labelling and transportation of specimens in section 7, due to withdrawal of procedure Ref IPC-0001-015 v3.	Agreed in principle at IPCC 19/01/23, pending retrospective final approval at IPCC April 20th 2023

Appendix 1 - Equality Analysis Screening Form

Please note; The Equality Analysis Policy and Equality Analysis Guidance can be found on the policy pages of the intranet

Name of Service area, Directorate/Department i.e. substance misuse, corporate, finance etc.	Nursing - Infection Prevention and Control					
Policy (document/service) name	CJD					
Is the area being assessed a…	Policy/Strategy		Service/Business plan		Project	
	Procedure/Guidance				Code of practice	
	Other – Please state					
Geographical area covered	Trustwide					
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 (This is the date you are asked to write or review the document/service etc.)	19/01/22					
End date of Equality Analysis Screening (This is when you have completed the equality analysis and it is ready to go to EMT to be approved)	21/01/22					

You must contact the EDHR team if you identify a negative impact - email tewv.eandd@nhs.net

1. Who does the Policy, Service, Function, Strategy, Code of practice, Guidance, Project or Business plan benefit?

2. Will the Policy, Service, Function, Strategy, Code of practice, Guidance, Project or Business plan impact negatively on any of the protected characteristic groups below?

Race (including Gypsy and Traveller)	No	**Disability** (includes physical, learning, mental health, sensory and medical disabilities)	No	**Sex** (Men, women and gender neutral etc.)	No
Gender reassignment (Transgender and gender identity)	No	**Sexual Orientation** (Lesbian, Gay, Bisexual and Heterosexual etc.)	No	**Age** (includes, young people, older people – people of all ages)	No
Religion or Belief (includes faith groups, atheism and philosophical belief's)	No	**Pregnancy and Maternity** (includes pregnancy, women who are breastfeeding and women on maternity leave)	No	**Marriage and Civil Partnership** (includes opposite and same sex couples who are married or civil partners)	No

Yes – Please describe anticipated negative impact/s

No – Please describe any positive impacts/s

Provides information on the care of patients with CJD for all people regardless of protected characteristics.

3. Have you considered other sources of information such as; legislation, codes of practice, best practice, nice guidelines, CQC reports or feedback etc.? **If 'No', why not?**	**Yes**	**yes**	**No**	

Sources of Information may include: • Feedback from equality bodies, Care Quality Commission, Equality and Human Rights Commission, etc. • Investigation findings • Trust Strategic Direction • Data collection/analysis • National Guidance/Reports	• Staff grievances • Media • Community Consultation/Consultation Groups • Internal Consultation • Research • Other (Please state below)

4. Have you engaged or consulted with service users, carers, staff and other stakeholders including people from the following protected groups?: Race, Disability, Sex, Gender reassignment (Trans), Sexual Orientation (LGB), Religion or Belief, Age, Pregnancy and Maternity or Marriage and Civil Partnership

Yes – Please describe the engagement and involvement that has taken place

IPC team reviews any case or suspected case of CJD across the trust and if identified will work will all staff through training and engagement to reduce the risk.

No – Please describe future plans that you may have to engage and involve people from different groups

5. As part of this equality analysis have any training needs/service needs been identified?					
Yes/No	Please describe the identified training needs/service needs below				
A training need has been identified for;					
Trust staff	Yes/No	Service users	Yes/No	Contractors or other outside agencies	Yes/No
Make sure that you have checked the information and that you are comfortable that additional evidence can provided if you are required to do so					

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	
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	