Trust Policy	Worcestershire Acute Hospitals NHS Trust

CREUTZFELDT–JAKOB DISEASE (CJD) AND VARIANT CJD (vCJD)

Department / Service:	Infection Prevention and Control
Originator:	Angela Roxburgh-Powell, Senior IPCN
Accountable Director:	Julie Booth, Deputy DIPC
Approved by:	Trust Infection Prevention & Control Committee
Date of approval:	4th May 2023
Review Date: This is the most current document and is to be used until a revised version is available	4th May 2026
Target Organisation(s)	Worcestershire Acute Hospitals NHS Trust
Target Departments	All Clinical Departments
Target staff categories	All Clinical Staff

Policy Overview:
This is an over-arching policy which provides guidance and advice around CJD and other transmissible spongiform encephalopathies (TSEs).

Key amendments to this guideline:

Date	Amendment	By:
May 2023	New Policy approved - replaces WAHT-INF-012 Creutzfeldt-Jakob Disease (CJD) and variant CJD (vCJD) – Minimising the risk of Transmission	TIPCC, Chaired by Jackie Edwards, interim CNO

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1. Introduction and Further Information

This policy provides advice on safe working practices with the aim of preventing the transmission of CJD and variant CJD (vCJD).

Further general advice is available from the UKHSA
https://www.gov.uk/government/publications/guidance-from-the-acdp-tse-risk-management-subgroup-formerly-tse-working-group

For IPC specific advice see:
https://assets.publishing.service.gov.uk/government/uploads/system/uploads/attachment_data/file/427854/Infection_controlv3.0.pdf

Or contact the trusts IPC team on 38752 or 44744.
Out of office hours please contact the on call microbiologist on duty, available via switchboard.

2. Definitions

The use of the term "CJD" in this policy encompasses sporadic CJD, sporadic fatal insomnia, variable protease-sensitive prionopathy (VPSPr), vCJD, iatrogenic CJD, genetic CJD, Fatal Familial Insomnia (FFI) and Gerstmann-Straussler-Scheinker Disease (GSS), in order to assist readability.

3. Responsibility and Duties

This guidance does not override the individual responsibility of health professionals to make appropriate decision according to the circumstances of the individual patient in consultation with the patient and /or carer. Health care professionals must be prepared to justify any deviation from this guidance.

4. What is CJD?

CJD is one of a group of diseases called transmissible spongiform encephalopathies (TSEs). These diseases are characterised by degeneration of the nervous system and are invariably fatal. CJD is the commonest of the human TSEs but it is still rare, in the UK about 60 cases are reported per year. The average age of onset of CJD is between 55 and 75 years, it has no known cause in the majority of cases. However, about 10% of cases are inherited and are caused by gene mutations. About 1% in the past have been transmitted as a result of medical treatments such as human pituitary derived growth hormone injections, corneal transplants and brain surgery involving contaminated instruments.

- **Symptoms of CJD**
    - Average age of onset – 60 years (range 16-83)
    - Rapidly fatal
    - Initial decline in attention, sleeping and eating patterns, memory and fatigue
    - Quickly develops into a rapidly progressive dementia
    - May be accompanied by aphasia, cortical visual failure, myoclonus, cerebellar ataxia, extrapyramidal features, prominent startle responses and late seizures
    - Unusual early features include vertigo and paraesthesia

5. What is variant CJD?

In 1996 a form of CJD was identified that differed from previously recognized types of the disease. The patients affected were younger, their symptoms were different and the appearance of their brain tissue after death was not the same as in CJD. This disease is known as variant CJD (vCJD). Analysis of the incidence data indicates that the vCJD epidemic reached a peak in mid-2000 and has since declined. The precise nature of the agent which causes vCJD is not known, but the most likely theory implicates an abnormal form of a protein which is called a 'prion'. The Government's Spongiform Encephalopathy Advisory Committee (SEAC) concluded that the most likely explanation for the emergence of vCJD was that it had been transmitted to people through exposure to Bovine Spongiform Encephalopathy (BSE).

- **Symptoms of vCJD**

  Distinguishing features from CJD are:

  - Slower clinical deterioration
  - Younger age of onset
  - Insidious onset of personality and behavioural change
  - Ataxia is more prominent

6. Key Points

- Follow Standard Infection Control precautions.
- Use disposable lumbar puncture sets.
- Instruments and equipment used in the care of patients with confirmed CJD should be disposed of by incineration.
- Instruments used on patients suspected of having CJD of any type should be quarantined pending confirmation of diagnosis.
- Flexible endoscopes must have a unique identifier recorded on every patient usage.
- Instruments and equipment used in procedures involving brain, spinal cord or eyes, carried out on a patient without CJD but in a risk category, should be destroyed by incineration.

7. Minimising the transmission risk of CJD

There is no evidence to suggest that CJD is spread from person to person by close contact, though it is known that transmission of CJD can occur in specific situations associated with medical interventions – **iatrogenic** infections. Due to the possibility of iatrogenic transmission of CJD, precautions need to be taken for certain procedures in healthcare, to prevent transmission.

CJD (except vCJD)
Worldwide, cases of iatrogenic CJD have been associated with the administration of hormones prepared from human pituitary glands and *dura mater* preparations, and one case has been reported associated with a corneal graft (it is possible that the corneal tissue was contaminated by posterior segment tissue during processing). Iatrogenic transmission has also been identified following neurosurgical procedures with inadequately decontaminated instruments or EEG needles.

vCJD
There have been no known transmissions of vCJD via surgery or use of tissues or organs. Since 2003, four cases (three clinical and one asymptomatic) of presumed person-to-person transmission of vCJD infection via blood transfusion of non-leucodepleted red blood cells have been reported in the UK. In addition, in 2009, a case of probable asymptomatic vCJD infection via plasma products was reported in a haemophiliac.

- **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
- patients “at increased risk”

In this policy document, the term ‘patients with, or “at increased risk” of, CJD’ is used as a proxy for all patient groups in Table 4a contained in the national guidance document https://assets.publishing.service.gov.uk/government/uploads/system/uploads/attachment_data/file/427854/Infection_controlv3.0.pdf Where this term is used, the guidance is applicable to all patient groups in Table 4a.

8. Tissue Infectivity, Surgical procedures, Instrument Management and Waste

For all patients with, or “at increased risk” of, CJD, the following precautions should be taken for surgical procedures:

- Wherever appropriate and possible, 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;
- Protective clothing should be worn, *i.e.* liquid repellent operating gown, over a plastic apron, gloves, mask and goggles, or full-face visor;
- for symptomatic patients, this protective clothing should be single-use and disposed of in line with local policies;
- for patients “at increased risk” of CJD, this protective clothing need not be single-use and may be reprocessed;
- 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 reusable instruments should be in place, so that instruments can be related to use on a particular patient.

For further information, please see **Appendix 1, 2 & 3**

- **Problems with surgical instruments**

If any problems are identified with instruments or sets of instruments, this should be referred to MHRA through the Yellow Card Scheme https://yellowcard.mhra.gov.uk/

- **Bed linen**

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

- **Childbirth**

Childbirth should be managed using standard infection prevention and control procedures. The placenta and other associated material and fluids are designated as low risk tissues, and should be disposed of as clinical waste, unless they are needed for investigation.

9. Occupational exposure

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.

Further Information can be obtained from section 4.41 in
https://assets.publishing.service.gov.uk/government/uploads/system/uploads/attachment_data/file/427854/Infection_controlv3.0.pdf

10. Endoscopy

Appendix 4 contains advice on the precautions to be taken for endoscopic procedures on patients with, or “at increased risk” of, CJD.

11. Ophthalmology

Appendix 5 contains advice on the precautions to be taken for ophthalmic procedures on patients with, or “at increased risk” of, CJD.

12. Anaesthesia and intensive care

See Infection Control in Anaesthesia at https://anaesthetists.org/Home/Resources-publications/Guidelines/Infection-prevention-and-control-2020

13. Dentistry

The risks of transmission of infection from dental instruments are thought to be very low provided satisfactory standards of infection prevention and control and decontamination are maintained. There is no reason why any patient with, or “at increased risk” of, CJD, should be refused routine dental treatment. Such people can be treated in the same way as any member of the general public.

Further information is available at https://www.england.nhs.uk/estates/health-technical-memoranda/

14. After death

Guidance on dealing with the bodies of patients with, or "at increased risk" of, CJD, is contained in Annex H.
https://assets.publishing.service.gov.uk/government/uploads/system/uploads/attachment_data/file/209766/Annex_H_-_After_death.pdf
This includes advice on carrying out post mortem examinations and transportation of bodies, and advice for undertakers on embalming, funerals and cremations.

15. Monitoring and Compliance

Not yet available

16. Plan for implementation

The Policy will be made available on the Trust Intranet site under policies – departments - IPC

17. Dissemination

The Policy will be made available on the Trust Intranet site under policies – departments - IPC

18. Policy Review

TIPCC will review this policy 3-yearly.

19. References

CREUTZFELDT–JAKOB DISEASE (CJD)AND VARIANT CJD (vCJD) – MINIMISING THE RISK OF TRANSMISSION	WAHT-INF- 012
Minimise transmission risk of CJD and vCJD in healthcare settings https://www.gov.uk/government/publications/guidance-from-the-acdp-tse-risk-management-subgroup-formerly-tse-working-group	
Infection Prevention and Control of CJD and variant CJD in Healthcare and Community settings https://assets.publishing.service.gov.uk/government/uploads/system/uploads/attachment_data/file/427854/Infection_controlv3.0.pdf	

20. Background

21. Equality requirements

No findings

22. Financial risk assessment

No impact

23. Consultation

Members of TIPCC will be consulted, representing all divisions.

24. Contribution List

This key document has been circulated to the following individuals for consultation;

Designation
Tracey Cooper DIPC
Emma Fulloway IPC Nurse Manager
Kerrie Howels Senior IPCN
Eve Neale Senior IPCN

This key document has been circulated to the chair(s) of the following committee's/ groups for comments;

Committee
TIPCC

25. Approval Process

The Trust Infection Prevention and Control Steering Group will approve this document.

26. Version Control

This section should contain a list of key amendments made to this document each time it is reviewed.

Date	Amendment	By:

Appendices

Appendix 1

Handling of instruments – patients with, or "at increased risk" of, CJD (other than vCJD)

Tissue Infectivity		Status of patient	
	Definite or probable	**Possible**	**At increased risk**
High* Brain Spinal cord Cranial nerves, specifically the entire optic nerve and the intracranial components of the other cranial nerves Cranial ganglia Posterior eye, specifically the posterior hyaloid face, retina, retinal pigment epithelium, choroid, sub retinal fluid and optic nerve Pituitary gland	Single use or Destroy or Quarantine for re-use exclusively on the same patient	Single use or Quarantine for re-use exclusively on the same patient pending diagnosis	Single use or Destroy or Quarantine for re-use exclusively on the same patient
Medium Spinal ganglia Olfactory epithelium	Single use or Destroy or Quarantine for re-use exclusively on the same patient	Single use or Quarantine for re-use exclusively on the same patient pending diagnosis	Single use or Destroy or Quarantine for re-use exclusively on the same patient
Low	No special precautions	No special precautions	No special precautions

Appendix 2

Handling of instruments – patients with, or "at increased risk" of vCJD

Tissue Infectivity		**Status of patient**	
	Definite or probable	**Possible**	**At increased risk**
High* Brain Spinal cord Cranial nerves, specifically the entire optic nerve and the intracranial components of the other cranial nerves Cranial ganglia Posterior eye, specifically the posterior hyaloid face, retina, retinal pigment epithelium, choroid, sub retinal fluid and optic nerve Pituitary gland	Single use or Destroy or Quarantine for re-use exclusively on the same patient	Single use or Quarantine for re-use exclusively on the same patient pending diagnosis	Single use or Destroy or Quarantine for re-use exclusively on the same patient
Medium Spinal ganglia Olfactory epithelium Tonsil Appendix Spleen Thymus Adrenal gland Lymph nodes and gut-associated lymphoid tissues	Single use or Destroy or Quarantine for re-use exclusively on the same patient	Single use or Quarantine for re-use exclusively on the same patient pending diagnosis	Single use or Destroy or Quarantine for re-use exclusively on the same patient
Low	No special precautions	No special precautions	No special precautions

Appendix 3

Disposal of clinical waste from patients with, or "at increased risk" of, CJD or vCJD

Diagnosis of CJD	High or medium risk tissue*	Low risk tissue and body fluids**
Definite	Incinerate	Normal clinical waste disposal
Probable	Incinerate	Normal clinical waste disposal
"At increased risk"	Incinerate	Normal clinical waste disposal

Appendix 4

Managing the Risk - Endoscopy

See Annex F at Annex F: Endoscopy (publishing.service.gov.uk)

Appendix 5

Managing the Risk – Ophthalmology

See Annex L at Microsoft Word - Annex L - Managing CJDvCJD risk in ophthalmology Updated Jan 2011.doc (publishing.service.gov.uk)

Appendix 6

Managing the Risk – Dentistry

Guidance on dealing with the bodies of patients with, or "at increased risk" of, CJD, is contained in Annex H.
https://assets.publishing.service.gov.uk/government/uploads/system/uploads/attachment_data/file/209766/Annex_H_-_After_death.pdf
This includes advice on carrying out post mortem examinations and transportation of bodies, and advice for undertakers on embalming, funerals and cremations.

Appendix 7

Algorithm chart for precautions for reusable instruments for surgical procedures on patients with, or "at increased risk" of, CJD, vCJD and other human prion diseases available on page 23 at https://assets.publishing.service.gov.uk/government/uploads/system/uploads/attachment_data/file/427854/Infection_controlv3.0.pdf

Supporting Document 1 - Equality Impact Assessment Tool
To be completed by the key document author and attached to key document when submitted to the appropriate committee for consideration and approval.

		Yes/No	Comments
1.	**Does the policy / guidance affect one group less or more favourably than another on the basis of:**		
	Age	No	
	Disability	No	
	Gender reassignment	No	
	Marriage and civil partnership	No	
	Pregnancy and maternity	No	
	Race	No	
	Religion or belief	No	
	Sex	No	
	Sexual orientation	No	
2.	**Is there any evidence that some groups are affected differently?**	No	
3.	**If you have identified potential discrimination, are any exceptions valid, legal and / or justifiable?**	No	
4.	**Is the impact of the policy / guidance likely to be negative?**	No	
5.	**If so can the impact be avoided?**	No	
6.	**What alternatives are there to achieving the policy / guidance without the impact?**	No	
7.	**Can we reduce the impact by taking different action?**	No	

Supporting Document 2 – Financial Impact Assessment

To be completed by the key document author and attached to key document when submitted to the appropriate committee for consideration and approval.

	Title of document:	Yes/No
1.	Does the implementation of this document require any additional Capital resources	No
2.	Does the implementation of this document require additional revenue	No
3.	Does the implementation of this document require additional manpower	No
4.	Does the implementation of this document release any manpower costs through a change in practice	No
5.	Are there additional staff training costs associated with implementing this document which cannot be delivered through current training programmes or allocated training times for staff	No
	Other comments:	