Mid Essex Hospital Services NHS
NHS Trust

Creutzfeld Jacob Disease or Variant Creutzfeld Jacob Disease	**Type: Clinical Guideline** **Register No: 04078** **Status: Public**

Developed in response to:	Health & Social Care Act 2008, Health & Safety Act 1974, COSHH 2002,
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Consulted With	Post/Committee/Group	Date
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Document Review History

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2.3 Healthcare Act – no previous versions can be located	Angela Hyman	24th January 2008
3.0	Sue Adams	September 2014
4.0	Sue Adams	7 June 2016

Index

1 Purpose

1.1 To comply with Health & Social Care Act 2008 and Care Quality Commission (CQC), as part of the registration, the trust must protect patients and healthcare workers who may be at risk of acquiring healthcare-associated infection (HCAI) including Creutzfeld Jakob Disease (CJD) and variant CJD (vCJD).

1.2 To provide guidance to staff on measures to prevent transmission of CJD or vCJD.

1.3 To provide guidance to staff regarding assessment of patients undergoing surgery and/or endoscopy.

1.4 To provide guidance to staff on the safe disposal and decontamination of surgical equipment.

1.5 To provide guidance to staff on safe management of spillages, bed linen and procedure after death.

2. Aim of the policy

2.1 The aim of this policy is to provide advice on safe working practices to all staff caring for these patients and to prevent transmission of CJD or vCJD and other human prion disease to other patients and staff.

3. Scope

3.1 This policy applies to patients 'symptomatic' and 'asymptomatic with an increased risk' of CJD or vCJD, including children.

4. Equality and Diversity

4.1 The Trust is committed to the provision of a service that is fair, accessible and meets the needs of all individuals.

5. Roles and Responsibilities

5.1 **Chief Executive**
The Chief Executive has overall responsibility for ensuring that the Trust has the necessary management systems in place to enable the effective implementation of this policy and overall responsibility for the health and safety of staff, patients and visitors

5.2 **Chief Nurse**
The Chief Nurse has strategic responsibility for ensuring systems are in place to facilitate nursing staff awareness of this policy and appropriate support is given to enable staff in delivering practice as outlined in this policy.

5.3 **Chief Medical Officer**
The Chief Medical Officer has strategic responsibility for ensuring systems are in place to facilitate awareness of this guideline and to ensure that appropriate support is given to enable medical staff in delivering practice as outlined in this policy.

5.4 **Director of Infection Prevention and Control (DIPC)**
The DIPC will have operational responsibility for the effective implementation of this policy.

5.5 **Infection Prevention and Control Team (IPT)**
The Infection Prevention and Control Team is responsible for ensuring all staff members are made aware of this policy.

5.6 **All staff**
All staff must comply with this policy and act in a responsible manner, liaising with the IPT in a timely manner if they need advice or support.

6. Introduction

6.1 Transmissible Spongiform Encephalopathies (TSEs) otherwise known as prion diseases are rare, fatal, degenerative diseases affecting the central nervous system, that occur in humans and certain other animal species. There are several recognised TSEs, including Creutzfeldt-Jakob disease (CJD) in humans, Bovine Spongiform Encephalopathy (BSE) in cattle and Scrapie in sheep. TSEs are thought to be caused by the build up of an abnormal form of the naturally occurring 'prion' protein in the brain.

6.2 The commonest human TSE is Creutzfeld-Jacob Disease (CJD). This occurs in various forms

- Sporadic CJD
- Variant CJD (vCJD)
- Familial CJD
- Iatrogenic CJD

6.3 Classic (sporadic) CJD occurs worldwide with a frequency of approximately one per million populations per annum.

6.4 Variant CJD (vCJD) is thought to have resulted from oral exposure to Bovine Spongiform Encephalopathy (BSE), probably through consumption of contaminated beef and beef products.

6.5 The abnormal prions proteins are present in certain tissues before patients show any symptoms and some carriers may remain asymptomatic. Consequently there is a risk of contamination of instruments used during invasive procedures such as surgery and endoscopy.

6.6 Prion are highly resistant to standard methods of disinfection and sterilisation, and therefore a special approach must be adopted in the care of patients, disposal of clinical waste and handling of surgical instruments and other medical device.

6.7 As non-CJD diseases are extremely rare this guidance will refer to CJD specifically. However the principles and guidance will be the same for other prion disease.

6.8 This policy is based on national guidance from the Advisory Committee on dangerous Pathogens Transmissible Spongiform Encephalopathy (ACDP TSE) Risk Management Subgroup: https://www.gov.uk/government/publication/guidance-from the-acdp-tse-risk-management-subgroup-fromerly-tse-working-group

7. Diagnosis and Treatment

7.1 Diagnosis is usually made clinically. There are currently no widely available laboratory tests for human CJD although the diagnosis can be confirmed by examination of the brain tissue after death. Brain biopsy may be used in investigating cases of suspected CJD but may not

be definitive in establishing a diagnosis. Reaching a clinical diagnosis of CJD may take time as some patients may have an atypical presentation. There are no proven specific treatments available.

7.2 All cases where CJD is suspected should be reported by the responsible clinician (usually a neurologist) to the National CJD Research and Surveillance Unit (NCJDRSU) in Edinburgh and to the local Public Health England Team so that any necessary action can be taken. Further details are available at http://www.cjd.ed.ac.uk/

7.3 Clinicians, ward and community staff caring for patients in whom the diagnosis of a CJD/vCJD is being considered or investigated are urged to contact the Infection Prevention and Control Team (IPCT) as soon as possible. In particular, it is imperative that once the diagnosis has been suggested clinical procedures using equipment intended for re-use on other patients are not carried out without prior discussion with IPCT.

8. Identification and categorisation of patient by risk

8.1. There is no effective test to identify carriers of prion disease. Risk prior to surgery/endoscopy is determined by a single question (all patients) or series of questions (for procedures involving high risk tissues)

8.2. When considering measures to prevent CJD or vCJD transmission to patients or staff, it is useful to make a distinction between symptomatic patients and asymptomatic patients

8.3. Symptomatic patients are those who fulfil the internationally accepted diagnostic criteria for definite, probable and possible CJD or vCJD

8.4. Asymptomatic patients are those with no clinical symptoms but who are potentially at risk of developing one of these diseases i.e. having a medical or family history which places them in one of the risk group

8.5. Categorisation of patients by risk is outlined in Appendix 1

9. Tissue infectivity

9.1. In additional to the risk categorization of the patient, the infectivity of the tissue being exposed / handled must be considered, as some material is classified as higher risk than others (See Appendix 2). This classification will inform the healthcare workers what additional requirements, if any, is needed to handle / dispose of reprocess equipment in contact with the affected patient

9.2. It is believed that the most infective material is obtained from the central nervous system and eye. Tissue classified as ‘high risk’ are:

- Brain
- Spinal cord tissue
- Posterior eye
- Optic nerve

9.3. Tissue with is thought to be less infective, and is classified as ‘medium risk’

- Olfactory epithelium
- Spinal ganglia
- Lymphoid tissue (vCJD)

9.4. Tissue with no evidence of infectivity are described as 'low risk', e.g. blood, saliva, body secretions, or excreta

10. Pre- assessment (Surgical and/or Endoscopy)

10.1 Identifying patients who are known, suspected or at increased risk of CJD, as well as the type of surgical procedure due to be carried out is key to the correct management of surgical instruments

10.2. All surgical and/or endoscopy patients must be risk assessed with regards to whether they are at increased risk of CJD or vCJD. This will be achieved by asking the following single question – '**Have you ever been notified that you are at an increased risk of CJD or vCJD for public health purpose'.** This question must be incorporated into the pre-assessment and admission documentation as appropriate, in order that potential risk is identified as early as possible

Patient's response	Action
No	Surgery or endoscopy should proceed using normal infection prevention and control unless the procedure is likely to lead to contact with high risk tissue.

Patient's response	Action
Yes	Surgery or endoscopy can proceed if it does not involve medium or high risk tissue. Please ask the patient to explain further the reason they were notified. Special infection prevention and control precautions should be taken for all surgery or endoscopy involving contact with medium or high infectivity tissue (see Appendix 3) and the local infection prevention and control team should be consulted for advice.
Unable to respond	Surgery or endoscopy should be proceed using normal infection prevention and control unless the procedure is likely to lead to contact with high risk tissue.

11. Additional recommendations for surgery and neuro-endoscopy which may involve contact with high risk tissue (Appendix 2)

11.1. Additional recommendations are only applicable to those assessing patients in neurosurgical and ophthalmic surgical departments for intradural and posterior ophthalmic surgical procedure. With regards to endoscopy, these additional recommendations are only applicable to those assessing patients for intradural neuro-endoscopic procedure

11.2. As well as asking all patients whether they have been notified as being at increased risk of CJD/vCJD, clinicians assessing patients **for procedures that involve contact with high risk tissue** should ask supplementary questions (as outlined in Appendix 3 and actions to be taken following questions in Appendix 4) to assess further their CJD/vCJD risk. If a patient has answered '**yes'** to the question '**Have you ever been notified that you are of**

CJD/vCJD?' there is no additional need to ask the questions in Appendix 3 as the patient's risk status has been established.

11.3. If a patient has been identified as potential risk and is having a procedure involving high risk or medium risk tissue, **the surgical team must discuss this as far in advance of the planned surgical procedure as possible.** Timely communication will allow additional information to be gathered and enable all relevant members of the teams, including the relevant decontamination provider, to ensure that appropriate disposal equipment is available or the correct measures are put in place for the management of reusable instruments appropriate.

12. Invasive procedures (Surgery & Endoscopy)

12.1. Patients that are classed **either** as symptomatic (possible, probable or definite) or at risk of CJD/vCJD, must be identified prior to any surgical or endoscopic procedures. (See Appendix 5)

12.2. In routine clinical contact, no additional precautions are needed for the care of patient in all risk groups

12.3. However, when certain invasive interventions are performed, there is potential for exposure to the agents of TSEs. In these situations it is essential that control measures are in place to prevent the iatrogenic transmission of TSEs

13. General measures for invasive procedures

13.1 Procedures on such patients, and the practicalities of instrument handling, storage, cleaning and decontamination, or disposal, should be planned carefully in advance

13.2. **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.**

13.3. Body secretion, 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 the samples taken or procedure performed will be low risk.

13.4. Contact with small volumes of blood (including inoculation injury) is considered low risk; however it is known that transfusion of large volumes of blood and blood components may lead to vCJD transmission.

13.5. 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 prevention and 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.

13.6. When taking biopsy specimens of medium or high risk tissue, e.g. tonsil biopsy in a patient with suspected vCJD, or intestinal biopsy in a patient at increased risk of vCJD every effort should be taken to minimise the risk of infecting the staff or contaminating the environment.

13.7. Samples from patients with or "at increased risk" of CJD/vCJD should be marked with a 'Biohazard' label, and **must not be** sent to be laboratory through the pod system, but taken by hand. It is advisable to inform the laboratory in advance that a sample is being sent.

13.8. In all cases to minimise the loss of instruments, single-use disposal instrument should be used whenever possible, but only if this does not affect the quality of the care.

13.9. All staff directly involved in procedures on patients in the risk groups, or in the subsequently re-processing or disposal of potentially contaminated items, should be aware of the specific precautions, and adequately trained.

13.10. Sufficient notice should be allowed for the necessary preparations, which should include informing the Decontamination Provider.

14. Precautions during invasive procedures (surgery and endoscopy) on definite, probable, possible, or at risk patients

14.1. Wherever appropriate and possible, the intervention should be performed in an operating theatre and the procedure should be performed at the end of the theatre list to allow normal cleaning of the theatre before the next session. If a procedure is performed on a ward it should take place in the treatment room. Involve only the minimum number of staff required (see Appendix 6).

14.2. The following protective clothing should be worn:

- Liquid-repellent operation gown, over a plastic apron
- Gloves
- Mask
- Visor or goggles

14.3. This protective clothing should be treated as single-use disposed of by incineration after use.

14.4. Instruments (single-use or otherwise) and other medical devices should be managed in line with ACDP TSE guidance.

15. Quarantining of Surgical Instruments

15.1. The user, the individual's clinician, the Decontamination Provider and the IPCT are all jointly responsible for ensuring the correct instruments/endoscopic equipment are identified quarantine, reprocessing and subsequent storage in a safe and secure place (see Appendix 6 & 7)

15.2. The final decision on whether to quarantine instruments/endoscopic equipment and on release for the use or incineration must be made by a senior member of IPC staff

15.3. Further information is also available from the ACDP TSE guidance – https://www.gov.uk/government/upload/syste,/attachment_data/file/209764/Annex_-_Quarantining_of_surgical_instruments.pdf

15.4. A form should be attached to the quarantined instruments – see Appendix 9

15.5. Please contact Hospital Sterilization and Decontamination Unit (HSDU) for advice on the arrangements for access to boxes and cupboards used for quarantine.

16. Surface decontamination and management of spillages

16.1. The infectious agents associated with TSE are unusually resistant to inactivation by chemicals and other processes. The removal of contaminating material and thorough cleaning of the surface are the most important elements in cleaning up spillages on a hospital ward.

16.2. Standard methods should be followed to clear up spillages on the ward, including spillages of blood and CSF, where gloves and apron should be worn. The spillages should be covered with paper towels to absorb the fluid then disposed of as clinical waste. All affected area regardless whether it is affected by medium to high risk materials should be disinfected with Tristel with minimum 5 minutes contact time following paper towel absorption.

17. Collection of laboratory samples

17.1. Body secretions and body fluids (including saliva, blood and CSF) are considered low risk for CJD/vCJD. It is therefore likely that the majority of samples taken and procedures performed on the ward will be low risk (also referred to Section 13.5 to 13.7).

17.2. The collection of all samples should involve standard precautions (i.e. avoidance of sharps injuries and other forms of parenteral exposure) and the safe disposal of sharps and contaminated waste by incineration. The procedure should be carried out by competent staff who are aware of the hazards involved. Lumbar puncture should only be carried out by competent staff who are aware of the hazards involved. Disposal gloves and eye protection should be worn. All lumbar puncture on any patient (regardless of a possible diagnosis of CJD or vCJD) must always be carried out with single-use lumbar kits.

17.3. **Must first discuss with Microbiology Laboratory before undertaking any procedures such as taking biopsy and CSF samples from cases of potential CJD. These samples should be marked with a 'Biohazard' on the request form.**

18. Laboratory handling of samples

18.1. The agents of TSE are classed as hazard group 3 pathogens, and all clinical specimens from definite, probable, possible or at risk patients should be handled at containment level 3. However, the option of derogation does not apply and, based on local risk assessment, certain containment level 3 precautions can be dispensed with.

18.2. All Trust laboratories must ensure that appropriate risk assessments have been made and that procedures are in place for safe handling of specimens from TSE patients. Such procedures must be applied to all specimens from definite, probable, possible, or at risk patients, and must include procedures for the inactivation and safe disposal of clinical specimens.

18.3. If samples are being sent to the National CJD Research Unit, this should be arranged directly with them, including courier pick-up. If the sample is to be picked up from a Trust laboratory, the laboratory needs to be informed that the sample is coming and the courier is due to collect it.

18.4. More detailed information can be found in the ACDP TSE Guidance Annex K – https://www.gov.uk/government/uploads/system/attachment_data/file/209769/Anne K_-_Guidelines_for_pathologist_and_pathology_laboratories.pdf

19. General care of CJD/vCJD in Hospital and Clinic Settings

19.1. The methods outlined below (Section 20 & 21) are applicable to patients with or at risk of CJD/vCJD unless otherwise stated.

19.2. There is no evidence of a risk to staff, relatives or the community from normal social or routine clinical contact with patients with or at risk of CJD/vCJD.

20. Isolation

20.1. For routine clinical contact with patients known or suspected to be suffering from CJD or vCJD continue to use Standard Infection Prevention and Control Precautions. Isolation (barrier nursing) is **not necessary** and the patient can be nursed on an open ward using standard precautions.

21. Bed linen

21.1. No additional handling or processing requirements are necessary soiled/contaminated linen should be handled in the steps laid out on the linen poster which should be displayed in all wards / departments.

22. Waste management

22.1. High or medium risk tissues (Appendix 2) and objects contaminated with high or medium tissues should be bagged or placed in a sharps bin (as appropriate) and sent for incineration in accordance with the Trust waste policy

22.2. Normal procedures apply for low risk waste

22.3. In the ward setting the majority of the clinical waste will be low risk and can be disposed of according to standard Trust policy. High or medium risk tissues (Appendix 2) and contaminated with high or medium risk tissues should be bagged or places in a sharps bin (as appropriate) and sent for incineration in accordance with Trust waste Management Policy. Special collection for incinerated waste must be arranged by individual department.

23. Childbirth

23.1. In the event that a symptomatic or at-risk patient is found to be pregnant, childbirth should be managed using standard infection prevention control precautions. The placenta, other associated materials and fluids are regarded as low risk and can be handled as per Trust policy.

24. Procedure after death

24.1. The IPCT must be informed of the death of a definite, probable, possible or known at-risk patient. The removal of the body from the ward to the mortuary should be carried out using normal infection control measures.

24.2. 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 the face, need not be discouraged, but **must occur prior to post mortem examination**.

25. Organ transplants

25.1. To minimise the risk of transmission of CJD/vCJD, organ donations should be rejected from patients with definite, probable or possible CJD/vCJD; those with degenerative neurological conditions of unknown cause and patients classified as ‘at-risk’ from CJD/vCJD.

26. Needlestick injuries

26.1. Any inoculation injury (needlestick injury) should be handled according to Trust Safe Handling and Disposal of Sharps policy, including informing the Occupation Health department. Contact with small volume 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.

27. Post mortems

27.1. Post mortem should always be avoided if all possible. If not, it is recommended that post mortem is only carried out in a high risk post mortem facility to reduce the risk of potential infection.

27.2. More detailed information can be found in the ACDT TSE Guidance Annex H. Specific guidance for the further management of the deceased in relation to undertakers and embalmers is also detailed in Annex H – https://www.gov.uk/government/uploads/system/uploads/attachment_data/file/209766/Annex_H_-_After_death.pdf

28. Training

28.1. These guidelines refer to all staff caring for patients with or at increased risk of TSE disease. The infection prevention team should be informed of all admissions to MEHT (to include emergency and elective admissions) to ensure appropriate advice and training is given.

28.2. All healthcare staff conducting pre-surgery assessment should receive instruction and/or training necessary to understand the reasons for asking these questions. This training should form part of the department’s induction programme.

28.3. It is important that these questions are asked in a manner that does not cause undue anxiety, and therefore the staff should be prepared and able to reassure the patient, and provide further information if needed.

29. Audit and monitoring

29.1. The Director of Infection Prevention & Control will formally review each case to include an audit of whether the guidance was followed in each case.

29.2. The medical records of patients with CJD will be retained as they are likely to be of ongoing scientific interest.

29.3. The Medical Records for patients with CJD to be annotated as follows:

- fix an ALERT sticker on the front cover of the records
- clinical staff to fill in the Alert information on the inside front cover of the records

- at the top of the inside front cover, not in a box write in 1inch capital letters “for permanent retention”

30. Implementation and communication

30.1. This guideline will be issued to the following staff groups to disseminate and ensure their staff are made aware of the guideline:

- Medical Director – issue to all Consultants and relevant junior doctors
- Heads of Nursing – issue to the relevant Lead nurses
- Ward Senior Sisters/Charge nurse – issue to relevant nursing staff within their ward
- Departmental Managers - issue to relevant staff within their department

30.2. The guideline will also be issued via the Staff Focus and made available on the Intranet and a hard copy available in the Ward/Department Infection Prevention Policy folder.

31. References

Department of Health (DH) 2013 ‘Minimising transmission risk of CJD and vCJD in healthcare setting’ https://www.gov.uk/government/publications/guidance-from-the-acdp-tse-risk-management-subgroup-formerly-tse-working-group

The Association of Anaesthetists of Great Britain and Ireland (AAGBI) 2008 ‘Infection control in anaesthesia’ http://www.aagbi.org/sites/default/files/infection_control_08.pdf

National Creutzfeldt - Jakob disease Surveillance Unit, Neuropathology Laboratory, western General Hospital, Crewe road, Edinburgh EH4 2XU. Telephone 0131 332 2117; fax 0131 343 1401 (http://wwwcjd.ed.ac.uk)

National Institute for Health and Clinical Excellence, Patient safety and reduction of risk of transmission of Creutzfeldt-Jacob disease (CJD) via interventional procedures November 2006. (http://www.nice.org.uk

Appendix 1

Categorisation of patients by risk

	Patient groups
Symptomatic patients	• Patients who fulfil the diagnostic criteria for definite, probable or possible CJD or vCJD (see Annex B of DOH TSE Guidance for diagnostic criteria – https://www.gov.uk/government/up;oad/system/uploads/attachment data/file/2099761/Annex B - Diagnostic criteria.pdf • 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
Asymptomatic 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 two or more blood relatives affected by CJD or prior disease
Asymptomatic patient identified as "at increased risk" of CJD/vCJD through iatrogenic exposures	• Recipients of hormone derived from human pituitary glands, e.g. growth hormone, gonadotropin, are "at increased risk" of transmission of sporadic CJD. In the UK the use of human derived gonadotropin was discontinued in 1973 and use of cadaver-derived human growth hormone was banned in 1985, **but** the use of these products may have continued in other countries after these dates • Individuals who underwent intradural brain or intradural spinal surgery 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. Patient 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" 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 1980 • Individuals who have given blood to someone who went on to develop vCJD; • Individuals who have received blood from someone who has also given blood to a patient who went on to develop vCJD; • Individuals who have been treated with certain implicated UK sourced plasma products between 1990 and 2001

	<u>Note</u> **A number of patients will have also been identified as 'at increased risk' by the CJD Incidents panel, for example due to having received blood from someone who later went on to develop vCJD. These patients and their GPs will normally have been notified of their status. As from 1st April 2013 the CJD Incidents panel will be disbanded. All CJD and related incidents will be managed by the local Health Protection Agency/Public Health England and the Infection Prevention and Control Team.**

Appendix 2

Tissues infectivity

The table below presents current information on the distribution of infectivity in tissue and body fluids in CJD & vCJD, based on data from experimental studies, where available, and on information from other studies of natural TSE disease in humans and animals.

Tissue	**CJD** Assumed level of infectivity	**vCJD** Assumed level of infectivity
Brain	High	High
Spinal cord	High	High
Cranial nerves (entire optic nerve and intracranial portions of others)	High	High
Cranial ganglia	High	High
Posterior eye	High	High
Olfactory epithelium	High	Medium
Spinal ganglia	Medium	Medium
Tonsil	Medium	Medium
Gut associated lymphoid tissue	Low	Medium
Appendix	Low	Medium
Spleen and thymus	Low	Medium
Other lymphoid	Low	Medium
Dura mater (reclassified December 2010)	Low	Low
Anterior eye and cornea	Low	Low
Peripheral nerve	Low	Low
Skeletal muscle	Low	Low
Dental pulp	Low	Low
Gingival tissue	Low	Low
Blood and bone marrow	Low	Low
CSF	Low	Low
Placenta	Low	Low
Urine	Low	Low
Other tissue	Low	Low

Supplementary questions for clinicians to assess further risk of CJD/vCJD

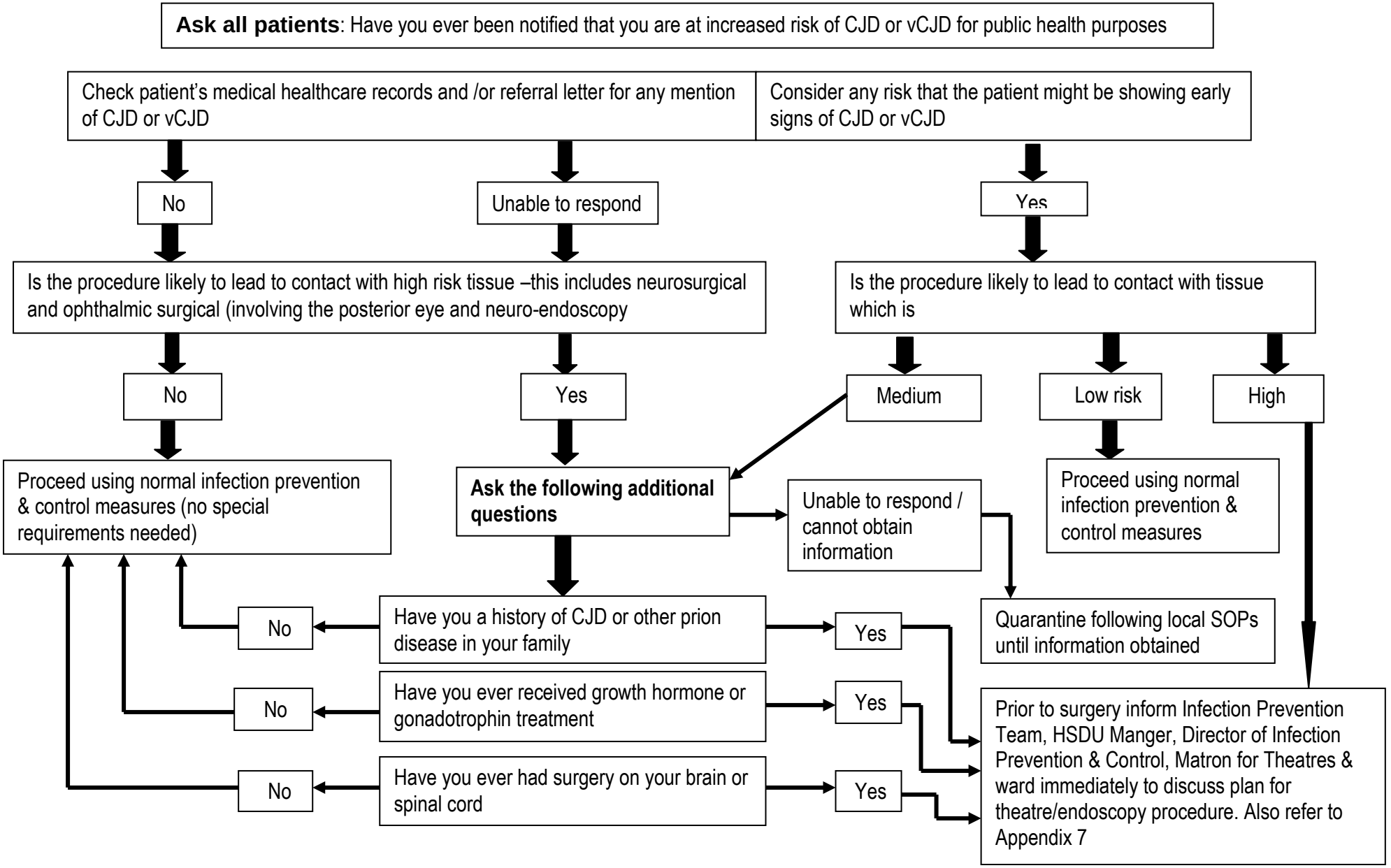
	Question to Patient	Notes to clinician
1	Have you a history of CJD or other prion disease in your family? If yes, please specify.	Patients should be considered to be risk from genetic forms of CJD if they have or have had: i) Genetic testing, which has indicated that they are at significant risk of developing CJD or other prion disease; ii) A blood relative known to have a genetic mutation indicative of genetic CJD or other prion disease; iii) 2 or more blood relatives affected by CJD or other prion
2	Have you ever received growth hormone or gonadotropin treatment? If yes, please specify: i) whether the hormone was derived from human pituitary glands ii) the year of treatment iii) whether the treatment was received in the UK or in another country	Recipients of hormone derived from human pituitary glands, e.g. growth hormone or gonadotropin, have been identified as at increased risk of sporadic CJD. In UK, the use of human-derived growth hormone was discontinued in 1985 but human-derived products may have continued to be used in other countries. In UK, the use of human-derived gonadotropin was discontinued in1973 but may have continued in other countries after this time.
3	Have you ever had surgery on the brain or spinal cord?	(a) Individuals who underwent intradural brain or intradural spinal surgery before August 1992 who received (or might have received) a graft of human-derived dura mater are “at increased risk” of transmission of sporadic CJD (unless evidence can be provided that human-derived dura mater was not used). (b) NICE guidance emphasises the need for a separate pool of new neuroendoscopes and reusable surgical instruments for high risk procedures on children born since 1st Jan 1997 and **who have not previously undergone high risk procedures**. These instruments and neuroendoscopes should not be used for patient born 1st Jan 1997 or those who underwent high risk procedures using reusable instrument before the implementation of this guidance.

Actions to be taken following questions in Appendix 3

Patient's response	Action
No to **all** questions	Surgery or neuro-endoscopy can proceed using normal infection prevention and control procedures.
Yes to **any** of questions **1.2 or 3**	Further investigations into the nature of the patient's CJD risk should be undertaken, and the patient's CJD risk assessed. This assessment of CJD risk should be recorded in the patient's medical notes/CRS for future reference. **If the patient is found to be at increased risk of CJD or vCJD following investigation, or risk status is unknown at the time of the procedure, special infection prevention and control precautions should be taken for the patient's procedure including quarantining of instruments, and Inform Infection Prevention Team and the Decontamination Lead, Clinical Director Microbiologist/Virologist and the Senior Nurse for the area immediately to discuss plan for theatre/endoscopy procedure** Please refer to the appropriate ACDP TSE Annex which gives guidance on the treatment of patients with or at increased risk of CJD. If the patient is found to be at increased risk of CJD or vCJD staff should contact the Infection Prevention team. Further information can be found using the following link http://www.nationalprionclinic.org
Unable to respond	In the event that a patient about to have emergency surgery or neuro-endoscopy is physically or otherwise unable to answer any questions, a family member, or someone close to the patient (in the case of a child, a person with parental responsibility) should be asked the CJD risk questions as set out in **Appendix 2** prior to the surgery or neuro-endoscopy. If the family member, or someone close to the patient, is not able to provide a definite answer to the CJD risk questions, the surgery or neuro-endoscopy should proceed but all instruments should be quarantined following the procedure (see Annex E of this guidance for details on Quarantining- https://www.gov.uk/government/up;oads/systems/uploads/attachment_data/file/209764/Annex E - Quarantining of surgical instruments.pdf

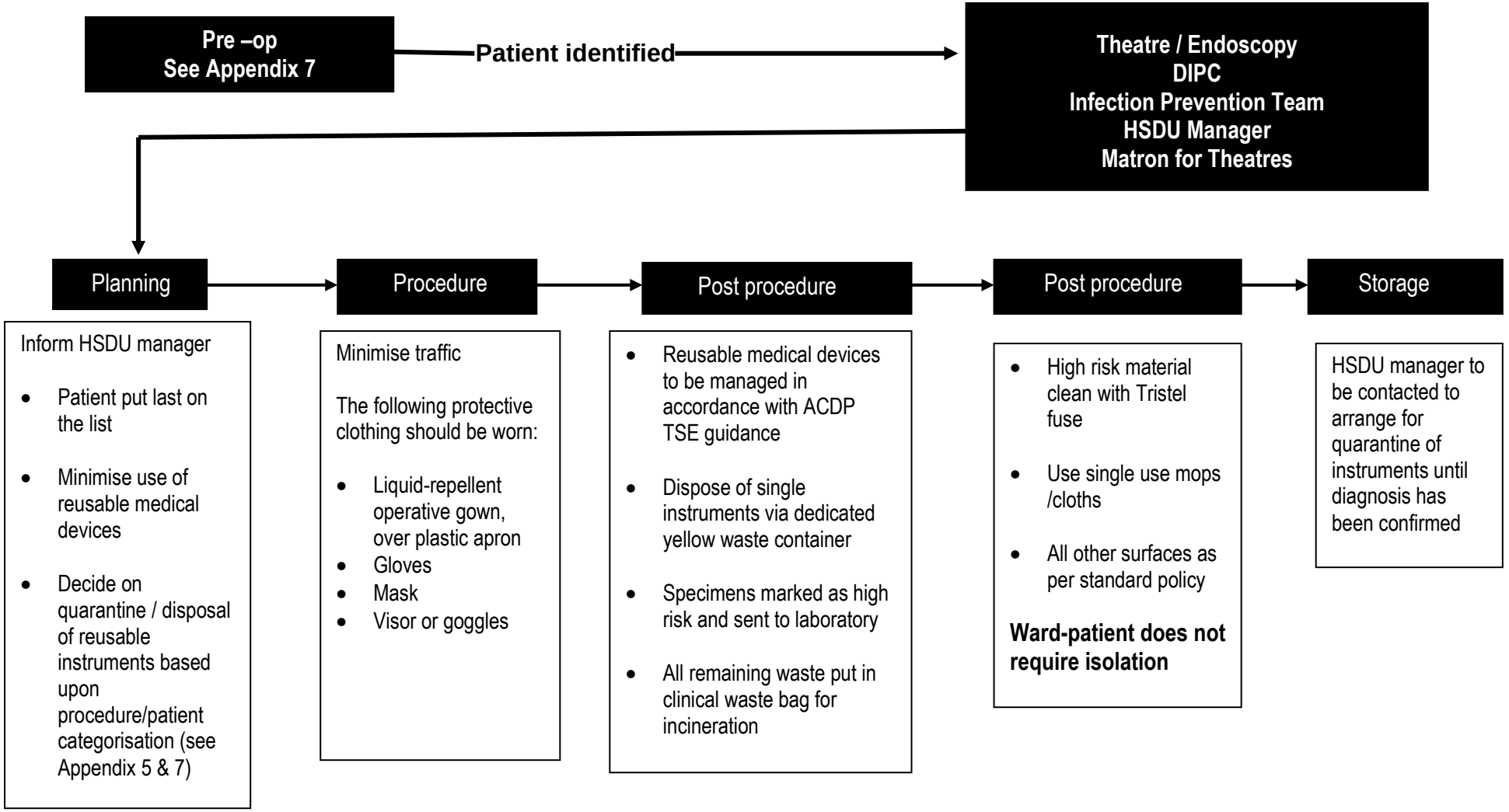
Appendix 5

Algorithm summarising CJD/vCJD risk assessment process for patients undergoing elective or emergency surgical or endoscopic procedures



Appendix 6

Elective theatre pathway for handling instruments on patients in high risk category for CJD / vCJD



Appendix 7

Actions to be taken with instruments following risk assessment for CJD/VCJD

1) Patients with symptoms of CJD or other TSE
   a) Definite or probable CJD/TSE diagnosis
   b) Possible CJD/TSE diagnosis

2) Patients at risk from inherited types of CJD but not showing any symptoms

3) Patients potentially at risk from exposure through medical procedures or treatment but not showing any symptoms

4) No risk patients (all patients not included in category 1-3 above)

Patient category	Tissue	Action
1a, 2 and 3	High or medium risk	Incinerate instruments after use
1a,2 and 3	Low risk (confirmed)	Reprocess
1b	High or medium risk	Test patient for prion disease. Quarantine instruments until test result received; incinerate if prion disease present or denote for single patient use; reprocess if prion disease not present.
1b	Low risk (confirmed)	Reprocess
4	High, medium or low risk	Reprocess

Appendix 8

Links to National Guidance form the **Advisory Committee on dangerous Pathogens Transmissible Spongiform Encephalopathy (ACDP TSE) Risk management Subgroup**

Annex A1	Distribution of TSE Infectivity in Human Tissues and Body Fluids	https://www.gov.uk/government/upload/system/uploads/attachment_ data/file/209761/Annex_B_-_Diagnostic_criteria.pdf
Annex B	Diagnosis of TSE"s	https://www.gov.uk/governmant/upload/system/uploads/attachment _data/file/209761/Annex_B_-_Diagnostic_criteria.pdf
Annex E	Quarantining of surgical instruments	https://www.gov.uk/government/uploads/system/uploads/attachment _data/file/209764/Annex_E_- _Quarantining_of_surgical_instruments.pfd
Annex F	Endoscopy (updated Jan 2013)	https://gov.uk/government/uploads/system/uploads/attachment_data/ file/270734/Annex_F_Endoscopy.pdf
Annex H	After death	https://www.gov.uk/government/uploads/system/uploads/attachment_ data/file/209766/Annex_H_After_death.pdf
Annex J	Assessment to be carried out before surgery and/or endoscopy (updated Jan 2013)	https://www.gov.uk/government/uploads/system/uploads/attachment _data/file/270735/Annex_J_Assessment_to_be_carried_out_before_ surgery_and_endoscopy_to_identify_patients_with_or_at_risk_of_CJD _vCJD.pdf
Annex K	Guidelines for pathologist and pathology laboratories for the handling of tissues from patients with, or at risk of, CJD or vCJD	https://www.gov.uk/government/uploads/system/uploads/attachment _data/file/209769/Annex_K_- Guidelines_for_pathologist_and_pathology_laboratories.pdf
Annex L	Managing vCJD risk in Ophthalmology	https://www.gov.uk/government/uploads/system/uploads/attachment_ data/file/209770/Annex_L_- _Managing_CJD_vCJD_risk_in_ophthalmology.pdf
Annex M	Managing vCJD risk in General Surgery and liver transplantation (new Jan 2013)	https://www.gov.uk/government/uploads/system/uploads/attachment_ data/file/209780?Annex_M_-_Managing_vCJD_risk.pdf
Part 4	Infection control of CJD, vCJD and other human prion diseases in healthcare and community settings (updated Jan 2013)	https://www.gov.uk/government/uploads/system/uploads/attachment_ data/file/270732/Part_4_Infection_Control_of CJD_vCJD_and_other_ human_prion_diseases_in_healthcare_and_community_settings.pdf

Appendix 9

Possible risk of CJD/vCJD Quarantine of instruments form

Patient Name:…….Hospital Number:…….

Date of Admission:……Hospital:……Ward:….....

Status of patient (definite, possible, probable, at risk)……

Date of procedure:……

Site:…….. Theatre/Endoscopy……..

Theatre/ Room Number……..

Name of the Lead Nurse…….. Surgeon/Doctor…….

Equipment quarantined:……

Date quarantined:…….

Equipment tracking number:….....

Quarantined dirty/clean: please stipulate (Please note that cleaning would only be applicable prior to quarantining endoscopes and only on the advice of the Infection Prevention and Control Team).

Infection Prevention Nurse (IPN) aware Yes/No

Name of IPN…… Date notified……..

Form completed By;

Name:…….Signature:…….

Date:……Time:…….

Please file original in notes; a copy with the container and a copy to be sent to Infection Prevention Team.

.....

Infection Prevention use only

Eventual Outcome: (Please indicate one of below)

1) Quarantine discontinue
2) Equipment sent for incineration
3) Equipment kept for same patient use……..Storage area…….

Reason for the above decision…….

Theatre/Endoscopy aware Yes/ No Date….....