Livewell Southwest

Transmissible Spongiform Encephalopathies (Prion diseases including CJD) and its Prevention in Healthcare Settings

Version No 3:6

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Assett Number:	**31**

Reader Information

Title	Transmissible Spongiform Encephalopathies (Prion diseases including CJD) and its Prevention in Healthcare Settings. V. 3.6
Asset number	31
Rights of access	Public
Type of paper	Policy
Category	Clinical
Document purpose/summary	To ensure that the risk of transmission of TSEs is minimised in the healthcare setting.
Author	Director of Infection Prevention & Control.
Ratification date and group	24th February 2016. Policy Ratification Group
Publication Date	3rd March 2016
Review date and frequency (one, two or three years based on risk assessment)	Three years after publication, or earlier if minor changes are required.
Disposal date	The Policy Ratification Group will retain an e-signed copy for the database in accordance with the Retention and Disposal Schedule; all previous copies will be destroyed.
Job title	Director of Infection Prevention & Control.
Target audience	All Staff.
Circulation	Electronic: Livewell Southwest (LSW) intranet and website (if applicable) Written: Upon request to the PRG Secretary on ☏ 01752 435104. Please contact the author if you require this document in an alternative format.
Consultation process	Dawn Slater. IPCC Committee.
Equality analysis checklist completed	Yes.
References/Source	See section 10.
Associated documentation	N/A
Supersedes Document	V3.4
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Document review history

Version no.	Type of change	Date	Originator of change	Description of change
V2.0		June 2005	Dr P Jenks	Nothing substantive.
V3.0	Updated	Dec. 2007	Dr P Jenks	Nothing substantive.
v3:1	Reviewed	Nov 2010	Inf Control Nurse Lead	Reviewed, no changes made.
V3:2	Reviewed	Dec 10	PRG	Review date extended, no other changes.
V3.3	Reviewed	Jun 2012	PRG	Review date extended, no other changes.
V.3.4	Reviewed	July 2012	Director of Infection Prevention & Control.	Updating to PCH. Template.
V.3.5	Extended	December 2014	Director of Infection Prevention & Control.	Extended no changes.
V.3.6	Reviewed	December 2015	Infection Prevention and Control Manager	Minor amendments

Transmissible Spongiform Encephalopathies (Prion diseases including CJD) and its Prevention in Healthcare Settings.

1. Salient Points and Summary

- Prevention of the hospital transmission of Transmissible Spongiform Encephalopathies (TSEs), including Creutzfeldt-Jakob Disease (CJD) and variant vCJD, depends on the identification of infected individuals with these diseases and those at risk of incubating disease. The Infection Prevention and Control Team (IPCT) should be informed as soon as such patients are identified. These individuals include:

    - Those with known or clinically suspected TSE
    - Those with dementia of unknown origin for which a diagnosis of TSE is being actively pursued
    - Those with a family history of TSE
    - Those who received a human dura mater graft prior to August 1992
    - Those who received human cadaveric pituitary hormones prior to 1986
    - Those who may have been contacted in the past with a TSE and informed that they are potentially at risk of TSE. These include those informed by Public Health England to be at 1% or greater potential additional risk of variant vCJD through treatment or transfusion with plasma products (including human albumin and immunoglobulin) made from plasma donated by people who later developed vCJD. All such individuals should have been informed of their respective exposures and the need to inform attending doctors in the future.

- Individuals who are at risk or with known or suspected TSE may be identified as :

    - Those already notified to the National CJD Surveillance Registry for known and suspected cases. This will be documented in the notes.

    - Certain patient at risk of vCJD will have a Patient Alert on their electronic and paper patient records.

    - By asking questions designed to identify those at risk of TSE. Sample questions are included in Appendix E.

- It remains the **responsibility of the admitting Doctor** to identify individuals with these conditions and to inform the relevant parties. It remains the responsibility of the **operating surgeon/physician** to ensure all operative instruments including endoscopes used on such individuals are dealt with as outlined in this policy. The IPCT and Sterilisation and Disinfection Unit (SDU)

manager are available to assist in the identification and management of individuals with TSE.

- Individuals with TSEs pose little risk to other patients and members of staff, providing Standard Infection Prevention Precautions are used.

- No patient at risk of or those with known or suspected TSE, including those with a neurological illness of unknown aetiology in whom a diagnosis of TSE is being considered, should undergo an invasive procedure or endoscopy without first discussing the individual with the IPCT and SDU Manager.

- Members of staff must undertake the appropriate precautions outlined in this document when performing invasive surgical procedures or endoscopy.

- The greatest risk to others occurs when surgical or endoscopy instruments used on affected individuals are reused. If individuals at risk of or those with known or suspected TSE, including those with a neurological illness of unknown aetiology in whom a diagnosis of TSE is being considered, have invasive procedures the surgical instruments used **may need to be destroyed**.

- If an inadvertent or emergency surgical or endoscopic procedure is performed on an individual at risk of, or with known or suspected TSE, including those with a neurological illness of unknown aetiology in whom a diagnosis of TSE is being considered, the used surgical instruments or endoscope should be placed in a secure site and the IPCT and SDU manager should be contacted immediately.

- Instruments used to biopsy non-focal Central Nervous System (CNS) lesions must either be single use or be quarantined until a diagnosis other than TSE is made. Due to logistical reasons the former option is preferred. If after the procedure TSE cannot be excluded and remains a possible diagnosis, the individual should be considered as a possible case of TSE and appropriate precautions followed.

2. Introduction

2.1 Description of Transmissible Spongiform Encephalopathies

Transmissible Spongiform Encephalopathies (TSEs) are rare neurological diseases characterised by a progressive and universally fatal course. The group of illnesses affect both humans and animals and in certain circumstances can cross from one species to another.

TSEs are unique as they are inheritable, infectious and able to arise spontaneously. The infectious agent is believed to be an abnormal form of a naturally occurring protein that is not readily degraded by enzymes, chemicals

or heat (1, 2). Standard steam sterilisation cycles are ineffective against these agents. The incubation period of a TSE may be years or decades.

2.2 Types of mammalian Transmissible Spongiform Encephalopathies

Human TSEs
Idiopathic: Sporadic Creutzfeldt-Jakob Disease (CJD) and Sporadic Fatal Insomnia
Inherited: Familial CJD, Fatal Familial Insomnia and Gerstmann-Straussler-Scheinker Syndrome
Acquired: Iatrogenic CJD, Variant (v) CJD and Kuru

Examples of Animal TSEs	
Bovine Spongiform Encephalopathy	Cattle
Scrapie	Sheep, Goats and Mouflon
Transmissible Mink Encephalopathy	Mink
Feline Spongiform Encephalopathy	Domestic and captive cats

2.3 Routes of transmission

Details of the human TSEs are given in Appendix A and either:

- are inherited from known and well defined familial lineages
- are acquired from reused contaminated surgical instruments
- are acquired from TSE contaminated tissues and drugs
- arise sporadically.

The overall incidence of TSE in the UK is one per million per year. Most sporadic disease occurs in the elderly with an incidence in this age group of 5 per million per year. Currently the majority of human disease is due to sporadic CJD. An as yet unquantified mode of transmission was from the ingestion of foodstuffs contaminated with bovine prions associated with Bovine Spongiform Encephalopathy (BSE; 'mad cow disease'). This route of transmission has resulted in the emergence of a new human clinical variant of CJD (vCJD).

Appendix A details the different forms of human TSE and the means of transmission. When considering measures to prevent transmission to patients or staff, it is useful to make a distinction between symptomatic patients, i.e. those who fulfil the diagnostic criteria for definite, probable or possible TSE, and asymptomatic patients i.e. 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 groups. Appendix B details the classification of the risk status of symptomatic and asymptomatic patients and Appendix C outlines diagnostic criteria.

3. Identification and reporting of individuals with TSE

3.1 Why it is important to identify individuals with TSE

It is important to identify individuals with TSEs because:

- Prion proteins are transmissible on surgical instruments
- Conventional sterilisation is ineffective at removing prion proteins
- Individuals incubating disease may be infectious whilst being asymptomatic
- Tissues other than the brain and spinal cord can be infectious

TSEs can be transmitted on surgical instruments. Those used directly on the CNS of symptomatic patients are of most risk. Operations on asymptomatic individuals incubating disease may also contaminate instruments leading to transmission of infection.

In vCJD there is extensive pre-symptomatic prion replication in peripheral lymphoid tissues, including tonsils, lymph nodes and gut-associated lymphoid tissue. These tissues represent potential sources of transmission of disease (2, 3).

Surgical instruments used on tissue potentially containing prion proteins must be destroyed by incineration. Clinical differentiation of the various clinical forms of TSE can be difficult so throughout this policy all TSEs are considered to have an equivalent potential to spread. It is important that individuals at risk of, or those with known or suspected TSE, including those with a neurological illness of unknown aetiology in whom a diagnosis of TSE is being considered and those having biopsy of non focal lesions of the CNS are identified pre-operatively to prevent the reuse of contaminated surgical instruments.

3.2 Actions to be taken on the initial diagnosis and on subsequent hospitalisations of an individual with known or suspected TSE

When individuals with TSEs are identified, the **IPCT and SDU Manager must be informed**. Although ante-mortem diagnostic techniques exist, the diagnosis is frequently made by post mortem histological examination of the brain.

All surgical instruments used on high/medium risk tissues (Appendix D) of patients individuals at risk of, or those with known or suspected TSE, including those with a neurological illness of unknown aetiology in whom a diagnosis of TSE is being considered, **must be destroyed**. Instruments must be placed in sealed containers and taken directly to the incinerator (contact IPCT/SDU for further details).

No patient individuals at risk of, or those with known or suspected TSE, including those with a neurological illness of unknown aetiology in whom a

diagnosis of TSE is being considered should have any form of endoscopic examination without first discussing it with the IPCT. If an inadvertent or emergency procedure is performed, place the used endoscope at a secure site and contact the IPCT.

If a diagnostic test for a TSE is requested for a patient, any surgical instruments used in that patient on high/medium risk tissue must be quarantined pending the test result and the IPCT informed.

3.3 Individuals at risk of incubating disease: Key points to elucidate when taking a patient history and action to be taken on identification of such patients

There is currently no pre-mortem test to identify individuals incubating disease. Certain individuals who are at a higher risk of incubating TSE must be identified pre-operatively and may even if asymptomatic pose a risk of subsequent transmission:

3.4 Familial history of TSE

Individuals who have two or more blood relatives affected by a TSE or they or a blood relative have documented evidence of a mutation associated with hereditary TSEs.

3.5 Receipt of a human dural mater graft prior to August 1992

3.6 Receipt of human cadaveric pituitary hormones prior to 1986

These will either be adults who received cadaveric Growth Hormone as children or women who received cadaveric Human Gonadotrophin.

3.7 Patients who have been contacted in the past and informed that they are potentially at risk of TSE.

Those who may have been contacted in the past with a TSE and informed that they are potentially at risk of TSE. These include those informed by the HPA to be at 1% or greater potential additional risk of variant vCJD through treatment or transfusion with plasma products (including human albumin and immunoglobulin) made from plasma donated by people who later developed vCJD. All such individuals should have been informed of their respective exposures and the need to inform attending doctors in the future.

3.8 Appendix E contains sample questions for use routinely when identifying whether an individual is at risk of incubating disease. In general, individuals in these risk categories are likely to know who they are.

3.9 If an individual falling into one of these groups is identified, the IPCT must be informed during the planning stages for any invasive procedure including

endoscopies. All surgical instruments used on high-risk tissues (Appendix D) **must be destroyed**. Instruments used in some operations involving medium risk tissues (Appendix D) may need to be destroyed and further information is available from the ICT. Where any doubt exists instruments should be destroyed. Instruments used on procedures involving only low risk tissues can be processed in the SDU as normal.

3.10 Actions to be taken prior to performing a surgical or endoscopic procedure on a patient with neurological disease of unknown aetiology where the diagnosis of TSE is being actively considered.

No surgical procedure should be performed on a patient with unexplained dementia until an opinion as to whether the patient may be suffering from a TSE is sought from a suitably qualified Specialist, such as a Consultant Neurologist or Neurosurgeon. If a diagnosis of TSE is likely, instruments used in surgical and procedures involving high/medium risk tissues (Appendix D) must be destroyed and endoscopy procedures carefully risk assessed. If an inadvertent or emergency procedure is performed place the used endoscope or surgical instruments in a secure site and contact the IPCT.

No patient at risk of or those with known or suspected TSE, including those with a neurological illness of unknown aetiology in whom a diagnosis of TSE is being considered, should undergo an invasive procedure or endoscopy without first discussing the individual with the IPCT and Sterilisation and Disinfection Unit (SDU) Manager.

4. General prevention of transmission of TSE in hospital

TSEs have rarely been transmitted from patient to patient. When it has occurred, transmission has been exclusively due to the reuse of neurosurgical instruments or the use of human hormones and contaminated tissue.

4.1 Instrument sterilisation

As the full burden of TSE in the community is not known all surgical instruments should be thoroughly cleaned and decontaminated, preferably in automated machines in the SDU. Thorough cleaning of instruments prior to decontamination is particularly important in reducing the risk of transmission of prion-associated disease. Failure to use automated methods must be supported with an adequate risk assessment and written Standard Operating Procedure (SOP). Anyone who washes an instrument by hand must be trained, wear appropriate personal protective equipment (PPE) and be vaccinated against hepatitis B.

4.2 General precautions

There is no reason to believe that horizontal transmission occurs outside of the specific conditions noted previously and basic infection control practices

are generally sufficient when managing potentially infected patients (6). With the exception of precautions relating to procedures outlined in Section 5, individuals at risk of incubating disease should be treated as for any other hospitalised patient.

The following precautions relate to those suspected or confirmed as having a TSE:

a. There is no need to isolate the individual for reasons other than those applicable to all patients.
b. The most infectious tissues such as cerebrospinal fluid (CSF) must be handled with care. Personal protective equipment, including gloves, apron and eye protection, should be worn for all procedures, including lumbar puncture (LP) and venepuncture. Only experienced staff should perform LPs and single use disposable equipment must be used (5).
c. Body secretions other than CSF are unlikely to be infectious for TSE and Standard Precautions should be used at all times.
d. Fouled bed linen should be handled as for all patients, as detailed in the Linen Services Policy.
e. Spillage of any bodily fluid should be mopped up by a suitably trained member of staff wearing appropriate PPE with an absorbent substance and the surface disinfected with bleach (1000 ppm available chlorine). All waste, including mop heads used to clean the spill area, towels and used PPE, should be placed in a thick (400-gauge) yellow clinical waste bag for incineration.
f. The spillage of a high/medium risk secretion or tissue (Appendix D) is likely to be an exceptional event. Initial cleaning should be with an absorbent material which should be disposed of by incineration. The contaminated surface should be disinfected with a bleach solution containing 20,000ppm available chlorine. This is a toxic and corrosive solution and must be used with great care. In this situation the IPCT should be informed and further advice sought.
g. Any inoculation injury or exposure of a mucous membrane to a secretion that may transmit a TSE (e.g. CSF) from a suspected, known or at risk individual should be dealt with as for any other inoculation injury (see Management of Inoculation Injuries Policy). The potentially infectious nature of the secretion should be noted on the incident form and sent to Occupational Health to ensure the exposure is formally documented.

4.3 Pregnancy

In the event of a woman with known or suspected TSE becoming pregnant the delivery should be managed using standard Infection Prevention and Control procedures. The placenta and other by-products of the birth should be considered infectious and disposed of as clinical waste unless required for investigations (see section 6). Instruments used during delivery can be reprocessed in the normal manner providing they have not been used on high or medium risk tissues (see Appendix D).

4.4 Staff Health and Wellbeing (SH&WB) notification of individuals potentially exposed to TSE

Individuals with TSEs pose little risk to other patients and members of staff providing Standard Precautions used with all patients are applied and appropriate precautions are taken when performing invasive surgical procedures.

The Department of Health have advised that a note should be made in the SH&WB files of staff who fall into one of the following categories and that the nature and risks of the exposure are discussed with them by the SH&WB:

- Any member of staff performing invasive clinical procedures on patients known or suspected to be suffering from a TSE, particularly where there is a risk of exposure to high and medium risk tissues (Appendix D)

- Laboratory staff handling tissue specimens from patients with known or suspected TSE

- Staff undertaking post-mortem examinations of patients who have died with a TSE

- Any inoculation injury should be managed according to the Management of Inoculation Injuries Policy. It is important that the Occupational Health Department is informed of the nature of the injury.

Responsibility for notifying SH&WB of individuals falling into these risk categories lies with the Consultant treating the patient for medical staff, the Senior Nurse of the Directorate treating the patient for nursing staff and the respective Heads of Departments for laboratory staff. Notification should occur for every contact.

A list of exposed individuals will be kept by SH&WB , the relevant contents of which will be available to the individual themselves and their Health and Safety Manager.

5. General infection control procedures for operative and endoscopic procedures on all patients at risk of TSE or at risk of incubating disease

5.1 Avoiding the reuse of TSE contaminated instruments

See Section 3 for what to do with instruments including endoscopes that have been used on any patient in an at-risk of TSE category.

In certain situations contaminated instruments may not be incinerated but donated for Prion research. This will be discussed on a case-by-case basis with the National CJD Surveillance Unit.

5.2 General planning

All surgical procedures should be carefully planned to allow all appropriate precautions and instrument selection to be made and minimising the number of instruments requiring incineration. Practicalities surrounding instrument cleaning and disposal need careful consideration. It ultimately remains the responsibility of the admitting doctor to identify individuals at risk of TSE and the operating surgeon to ensure the correct procedures as detailed in this document are followed. The IPCT and SDU manager are available to assist with this and should be involved at an early planning stage.

Non-invasive procedures do not require specific precautions over and above those needed for any other patient.

5.3 Anaesthetic Equipment

Single-use anaesthetic equipment should be used wherever possible. Any re-usable equipment used, for example in the maintenance of an airway in individuals at risk of, or those with known or suspected TSE, including those with a neurological illness of unknown aetiology in whom a diagnosis of TSE is being considered, should be destroyed or quarantined pending discussion with the IPCT. It is vital that fibre optic equipment is avoided unless one is clear that the exposure is such that destruction of the equipment is not warranted (see section 5.7).

5.4 Instrument Disposal

All instruments for incineration must be transported from the theatre to the incinerator in a sealed container. On the rare occasions that quarantining instruments is considered, the IPCT will advise on instrument storage. All other instruments may be processed as per normal protocol. It remains the responsibility of the operating surgeon to ensure that the instruments are delivered to the appropriate site (i.e. incinerator or SDU). A named member of the Theatre Team should be responsible for transportation of the instruments.

5.5 Precautions during invasive procedures

The risk of cross infection with TSE to the operating team is extremely low. In the absence of a needle stick injury there is probably no residual risk. There is no evidence of airborne transmission. It is however prudent:

a. To wherever possible perform invasive procedures in an operating theatre.

b. To perform the procedure at the end of the list to allow normal cleaning of theatre surfaces before the next session.
c. To involve the minimum number of personnel.
d. To maintain a one-way flow of instruments using single-use instruments.
e. For all individuals in the theatre to wear liquid repellent gowns over a plastic apron, gloves, mask, visor and goggles. These should be single-use only and destroyed by incineration.

5.6 Neurosurgical biopsy of non-focal CNS lesions and biopsy in patients with dementia of unknown cause

Brain biopsies are often performed for diagnostic purposes on patients with non-focal CNS lesions **and dementia of unknown cause.** Surgical instruments used on such patients should be considered potentially infectious with TSE and be destroyed by incineration (Appendix F). In certain circumstances instruments may be quarantined if a diagnosis can be made within a reasonable time frame (e.g. 1-2 weeks). The instruments will only be released if a definite diagnosis (other than TSE) can be made. On the rare occasions that quarantining instruments is considered, the IPCT will advise on instrument storage. If a definitive diagnosis cannot be made, consideration should be given to notifying the National CJD Research & Surveillance Unit of the case and instruments used in any subsequent invasive procedures destroyed.

5.7 Decontamination and handling of endoscopes.

The risk of transmission of prions during endoscopy is low with the exception of endoscopes used on central nervous tissue. Due to more extensive tissue deposition, vCJD poses a greater risk of endoscopic transmission (appendix D).

5.7.1. General prevention of unrecognised prion contamination

Normal cleaning reduces to a low level the risk of disease transmission following endoscopy. This risk can be further reduced for all endoscopic procedures by using:

- Single patient use channel cleaning brushes and instrument channel port valve.

- Single patient use biopsy forceps.

5.7.2. Specific endoscopic procedures

a) Central nervous system endoscopy

Introduction of an endoscope into the central nervous system (including the eye) should be avoided in individuals at risk of, or those with known or suspected TSE, including those with a neurological illness of unknown aetiology in whom a diagnosis of TSE is being considered. Where this is unavoidable, a single-use instrument should be used or the used instrument quarantined pending definitive diagnosis in those with suspected TSE.

b) Nasal Cavity endoscopy

Different endoscopic procedures have different risks of prion contamination. In patients in all risk groups, olfactory epithelium is considered a medium risk tissue (Appendix D). The olfactory epithelium is normally located deep within the nasal turbinates but its distribution varies between individuals and ectopic epithelium can be found throughout the cavity. This poses a special risk for endoscopic procedures involving the nasal cavity. No such procedure should be performed on an individual at risk of, or those with known or suspected TSE, including those with a neurological illness of unknown aetiology in whom a diagnosis of TSE is being considered, without considering the risks and benefits in detail with the IPCT and Endoscopist.

Introduction of an endoscope into the nasal cavity should be avoided wherever possible in individuals with known or suspected TSE, including those with a neurological illness of unknown aetiology in whom a diagnosis of TSE is being considered. If the procedure is essential, contact the IPCT. For patients with definite, probable or possible CJD, a single use endoscope should be used. If a nasendoscope does become contaminated with the olfactory epithelium, the endoscope should be removed from use and the IPCT informed immediately. The advice of the consultant carrying out the endoscopic procedure in the nasal cavity should be sought to determine whether a risk of contamination of the endoscope with olfactory epithelium can be excluded with confidence. If the procedure is performed inadvertently or as an emergency, quarantine the instrument in a secure location and contact the IPCT.

Individuals who are at risk of incubating disease may undergo safe nasal endoscopy if there is little risk of contact with the olfactory epithelium and a disposable sheath is used, which should then be destroyed by incineration. Following use on such a patient, normal decontamination procedures should be followed, unless there is evidence of failure (tears in the sheath or moisture on the nasendoscope). If failure is suspected, the endoscope should be removed from use and the IPCT informed immediately.

c) Other endoscopic procedures

All other non-neurological and non-nasal endoscopies can be safely performed on individuals in all groups **providing** no biopsies or invasive interventions are performed (as defined by Advisory Committee on Dangerous Pathogens). If the condition the patient is suffering from is NOT **v**CJD or is at risk of incubating a prion disease that is NOT **v**CJD then invasive procedures, such as taking biopsies, are safe and require no precautions over and above that for a standard procedure. Where any doubt exists contact the IPCT.

5.8 Request/Referral for TSE Diagnostic tests

If the diagnosis of TSE is considered likely enough to perform a TSE diagnostic test then until a diagnosis other than TSE can be made, the patient should be considered as a possible case. Instruments used on any surgical procedures involving medium or high risk tissues (Appendix D) must be destroyed (or on rare occasions quarantined). If a definitive diagnosis cannot be made consideration should be given to notifying the National CJD Surveillance Unit of the case.

6. Sample Collection and Labelling

6.1 Potential TSE Hazards

High and medium risk tissues (Appendix D) from a patient at risk of, or those with known or suspected TSE, including those with a neurological illness of unknown aetiology in whom a diagnosis of TSE is being considered should be considered a biohazard and samples only taken by appropriately trained staff. Samples should be sealed in robust containers and labelled as a ‘Danger of Infection’. The request form should discretely note the nature of the disease (e.g. note as ‘TSE’ and not ‘CJD’).

In all patient groups blood, urine, faeces, and superficial swabs pose a minimal risk of TSE transmission unless contaminated with high or medium risk tissue.

If a specimen is taken to diagnose a TSE the relevant laboratory must be warned in advance. The receiving laboratory must inform the IPCT. In general most specimens will be processed as normal though protein estimates will not be performed on CSFs. Culture of high and medium risk tissues will where possible be avoided.

Single use instruments must be used wherever possible when taking diagnostic samples and all instruments potentially contaminated with TSE destroyed.

6.2 Diagnostic laboratories

All laboratories dealing with human specimens must have their own individual Risk Assessments. This policy summarises the main features of dealing with specimens from patients at risk of or with known and suspected TSE.

6.2.1 Samples from individuals known or suspected of having a TSE or those with unexplained neurological disease where TSE is being actively considered

Wherever possible, equipment used on high or medium risk specimens from individuals known or suspected of having a TSE should be single use only and disposed of by incineration (for example, single-use counting chambers). Where this is not possible it is vital that the equipment is well maintained and regularly cleaned and disinfected.

6.2.2. Samples from individuals at risk of disease

Samples containing or contaminated with medium or high risk tissues (Appendix D) should be considered potentially contaminated with TSE and treated accordingly.

6.2.3. Neuropathology specimens

All preparations of nerve and brain tissue from at risk of, or with known or suspected disease should be treated as potentially infectious and handled in the laboratory at Category 3 (subject to appropriate derogation). Further information is available in Reference 2.

7. Procedures after death of individuals known suspected or individuals at risk of incubating disease

As with any deceased patient where there is a residual infectious risk the body should be placed in a body bag.

For advice regarding precautions to be taken by undertakers and embalmers please see the Policy for Handling Cadavers or contact the Consultant in Communicable Disease Control (number available from IPCT). In general viewing and hygienic preparation can be performed, but embalming is not recommended.

There is no reason why patients with confirmed TSE cannot be buried as opposed to cremated.

Post mortem examination of the brain is currently the only way of confirming the diagnosis of TSE and further advice regarding where and how the post mortem will be performed is available from the Department of Histopathology.

8. Checklist on what to do on identifying an individual at risk of, or those with known or suspected TSE, including those with a

neurological illness of unknown aetiology in whom a diagnosis of TSE is being considered.

A. Suspected or Confirmed Disease

Inform the IPCT and SDU manager that such an individual is in the hospital.

Ensure no invasive procedures are performed without first considering the guidance in this policy. If in doubt contact the IPCT or SDU manager.

If any procedure, such as an emergency operation, is performed before the IPCT or SDU manager can be informed, ensure all instruments used on high or medium risk tissues (Appendix D) are sent for disposal by incineration. If any doubt exists as to whether the instruments need to be destroyed place them in a sealed and labelled container and seek advice urgently from IPCT or SDU manager. Until the issue of instrument disposal is resolved the operating surgeon retains responsibility for the safe keeping of the instruments. This also applies to rigid and flexible endoscopes.

Ensure that the individual has been reported to the National CJD Surveillance Centre.

B. Neurological disease of unknown aetiology where TSE is actively being considered

Seek a neurology or neurosurgery opinion prior to planning any invasive procedure or endoscopy. If the diagnosis of TSE is being actively considered follow the relevant precautions outlined for a confirmed case.

C. At risk of Incubating Disease

Inform the IPCT and SDU manager that such an individual is in the hospital.

Ensure no invasive procedures are performed without first considering the guidance in this policy. If in doubt contact the IPCT or SDU manager. If any procedures, especially emergency ones, are performed before the IPCT or SDU manager can be informed, ensure all instruments used on high or medium risk tissues (Appendix D) are sent for disposal by incineration. Instruments used on other medium risk tissues, such as lymphoid tissue, may not need to be incinerated depending on the nature of the TSE the individual may be incubating. If a non **v**CJD disease is considered, quarantine the instruments in a secure place and contact the ICD/SDU Manager. If in doubt destroy all instruments exposed to high and medium risk tissues.

9. NICE Interventional Procedure Guidance 196

In November 2006, the National Institute for Health and Clinical Excellence (NICE), issued Interventional Procedure 196: ‘Patient safety and reduction of

risk of transmission of Creutzfeldt-Jakob disease (CJD) via interventional procedures'. The Trust is working towards full compliance with these recommendations which were as follows:

9.1. For high-risk surgical procedures (intradural operations on the brain and operations on the retina or optic nerve – 'high-risk tissues'):

- Steps should be taken to ensure that instruments that come into contact with high-risk tissues do not move from one set to another. Practice should be audited and systems put in place to allow surgical instruments to be tracked, as required by Clinical Framework for local Policy and Procedures 01-01 and 01-06:

  Supplementary instruments that come into contact with high-risk tissues should either be single use or should remain with the set to which they have been introduced.

9.2. All accessories used through neurendoscopes should be single use.

9.3. A separate pool of new neurendoscopes and reusable surgical instruments for high-risk procedures should be used for children born since 1 January 1997 and who have not previously undergone high-risk procedures. These instruments and neurendoscopes should not be used for patients born before 1 January 1997 or those who underwent high-risk procedures before the implementation of this guidance.

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All policies are required to be electronically signed by the Lead Director. Proof of the electronic signature is stored in the policies database.

The Lead Director approves this document and any attached appendices. For operational policies this will be the Locality Manager.

The Executive signature is subject to the understanding that the policy owner has followed the organisation process for policy Ratification.

Signed: Lead Nurse, Director of Infection, Prevention and Control

Date: 1st March 2016

Appendix A
Human TSE's and their means of transmission

Means of transmission	Examples of transmission	Comment
Surgical instruments	Classic CJD Theoretical risk from vCJD	Two cases of classic disease identified from the use of contaminated cerebral electrodes and five cases following neurosurgery.
Oral	vCJD Kuru	Uncertain, from hundreds to hundreds of thousands may be incubating disease. There have been 9-28 cases per year between 1995-2004. Ritualistic cannibalism; no evidence of transmission since the 1960s.
Blood products	vCJD	4 cases
Tissue transplants: Human dural graft Human corneal transplant Human pericardial graft, bone graft and liver transplant	 Classic CJD Classic CJD Classic CJD	 Over 114 cases. 3 cases. One case each. These are anecdotal cases the transplant cause of the CJD was never confirmed
Human hormone therapy: Growth Hormone Gonadotrophic hormone	 Classic CJD Classic CJD	 139 cases. 4 cases.
Inherited disease	Classic CJD, GSS, FFI	Rare
Sporadic disease	Classic CJD, GSS, FFI	1 per million per year

Appendix B

Categorisation of patients by risk

Patients should be categorised as follows, in descending order of risk:

1. Symptomatic patients	1.1 Patients who fulfil the diagnostic criteria for definite, probable or possible CJD or vCJD (See Appendix C for diagnostic criteria). 1.2 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.
2. Asymptomatic patients at risk from familial forms of CJD linked to generic mutations	2.1 Individuals who have been shown by specific genetic testing to be at significant risk of developing CJD or other prion disease. 2.2 Individuals who have a blood relative known to have a genetic mutation indicative of familial CJD. 2.3 Individuals who have or have had two or more blood relatives affected by CJD or other prion disease.
3. Asymptomatic patients identified as potentially at risk due to iatrogenic exposures ##	3.1 Recipients of hormone derived from human pituitary glands, e.g. growth hormone, gonadotrophin. In the UK, cadaver-derived human growth hormone was banned in 1985 but use of human-derived products may have continued in other countries. 3.2 Individuals who have received a graft of *dura mater*. (People who underwent Neurosurgical procedures or operations for a tumour or cyst of the spine before August 1992 may have received a graft of *dura mater*, and should be treated as *at risk*, unless evidence can be provided that *dura mater* was not used). 3.3 Patients who have been contacted as potentially at risk, including individuals considered to be **: a. at risk of CJD/vCJD due to exposure to

	certain instruments used on a patient who went on to develop CJD/vCJD, or was at risk of vCJD; b. at risk of vCJD due to receipt of blood components or plasma derivatives; c. at risk of CJD/vCJD due to receipt of tissues/organs d. at risk of vCJD due to the probability they could have been the source of infection for a patient transfused with their blood who was later found to have vCJD.

NB: A decision on the inclusion of corneal graft recipients in the '*iatrogenic at risk*' category is pending completion of a risk assessment

** The CJD Incidents Panel, which gives advice to the local team on what action needs to be taken when a patient, who is diagnosed as having, or found to be at risk of, CJD or vCJD, underwent surgery or donated blood, organs or tissues before CJD/vCJD was identified, will identify contacts who are potentially at risk.

Appendix C

Diagnostic criteria

Taken from Annex B of Transmissible Spongiform Encephalopathy Agents: Safe Working and the Prevention of Infection
https://www.gov.uk/government/uploads/system/uploads/attachment_data/file/260961/report.pdf

B.1 **Paragraphs** in Part 4 of this guidance, including Table 4, categorise CJD patients in descending order of risk, distinguishing between **symptomatic** and **asymptomatic** patients. **Symptomatic** patients are those who fulfill the internationally accepted diagnostic criteria, set out below, for **definite**, **probable** and **possible** CJD or vCJD.

B.2 Suspect cases are classified according to these criteria by a neurologist from the National CJD Surveillance Unit (NCJDSU), on an on-going basis. The classification is recorded at 4 "key" stages:

- at notification
- when the patient was first seen, in life, by a neurologist from the NCJDSU
- the highest classification on the sole basis of clinical information (i.e. not including neuropathological information)
- when a NCJDSU review is completed (i.e. when the case-file is closed). The completed case file may, or may not, include neuropathological data.

B.3 The date of any change of classification and the reason for such a change is recorded as necessary.

Classification criteria

Sporadic CJD

B.4 Neuropathological/immunocytochemical confirmation is required for a diagnosis of **definite** sporadic CJD.

B.5 **Probable** sporadic CJD patients will have rapidly progressive dementia, and at least two of the following four symptoms:

(a) myoclonus
(b) visual or cerebellar problems
(c) pyramidal or extrapyramidal features
(d) akinetic mutism

plus typical electroencephalogram (EEG) with generalised triphasic periodic complexes at approximately 1 per second,
or clinical criteria for **possible** sporadic CJD (see below) and a positive assay for 14-3-3 protein in the cerebrospinal fluid (CSF).

B.6 **Possible** sporadic CJD patients will have rapidly progressive dementia, two of the symptoms listed in paragraph B.5(a)-(d) above and a duration of less than 2 years.

Iatrogenic CJD

B.7 Iatrogenic CJD patients display progressive cerebellar syndrome in a pituitary hormone recipient or sporadic CJD with a recognised exposure risk (e.g. *dura mater* transplant) – see Part 4, Table 4a, section 3 of this guidance. A definite diagnosis of iatrogenic CJD still require a neuropathological examination.

Familial CJD

B.8 Patients with **familial** CJD will have **definite** or **probable** CJD (see definitions in paragraphs B.4 and B.5 above), plus **definite** or **probable** CJD in a first degree relative (i.e. a parent, child or sibling) or a neuropsychiatric disorder plus a disease-specific mutation in the prion protein gene.

Variant CJD (vCJD)

B.9 **Definite** vCJD patients will have a progressive neuropsychiatric disorder and neuropathological confirmation of the disease, showing spongiform change and extensive PrP C deposition with florid plaques throughout the cerebrum and cellebellum.

B.10 **Probable** vCJD patients can be classified under two sets of criteria:

(i) They will have progressive neuropsychiatric disorder of a duration greater than 6 months where routine investigations do not suggest an alternative diagnosis. They will also have at least four of the following five symptoms:

(a) early psychiatric symptoms (depression, anxiety, apathy withdrawal, delusions)
(b) persistent painful sensory symptoms (including both frank pain and/or unpleasant dysaesthesia)
(c) ataxia
(d) myoclonus or chorea or dystonia
(e) dementia

An EEG will not show the typical appearances of sporadic CJD, or no EEG has been done and there is a symmetrical high signal in the posterior thalamus on a MRI brain scan (1 Zeidler *et al* 2000).

These patients would have had no history of potential **iatrogenic** exposure.

(ii) Alternatively, a **probable** vCJD patient will have had a progressive neuropsychiatric disorder for a period of longer than six months, where routine investigations do not support an alternative diagnosis, and where there is no history of potential of iatrogenic exposure, plus a positive tonsil biopsy, which is positive for PrP-res.

B.11 **Possible** vCJD patients will have progressive neuropsychiatric disorder of a duration greater than 6 months where routine investigations do not suggest an alternative diagnosis, and no history of potential iatrogenic exposure. They will also have at least four out of five of the symptoms listed in paragraph B.10(I)(a)-(e) above and an EEG does not show the typical appearance of sporadic CJD or no EEG has been performed.

Patients who do not fulfill the criteria for possible CJD

B.12 The NCJDSU have designated three additional categories for patients who are referred to the Unit but who do not meet the criteria for **possible** CJD. These can be summarised as:

(i) **Diagnosis unclear** –the diagnostic criteria for **definite**, **probable** or **possible** CJD are not met, nor is there a reasonable alternative diagnosis. CJD, therefore, remains a possibility;

(ii) **CJD thought unlikely** – information indicates that a clinical diagnosis of CJD is very unlikely because of atypical disease features, and/or an atypical course, and/or atypical clinical investigation results, and/or a reasonable alternative diagnosis is made but is not confirmed. This category includes cases, which recover clinically without a firm alternative diagnosis;

(iii) **Definitely not CJD** – information indicates that CJD is not the diagnosis and there is an alternative definite diagnosis proven on the basis of clinical examination, clinical investigations or pathology.

Zeidler M, Sellar RJ, Collie DA, Knight R, Stewart G, Macleod MA, Ironside JW, Cousens S, Colchester AC, Hadley DM, Will RG. The pulvinar sign on magnetic imaging in variant Creutzfeldt- Jakob Disease. Lancet. 2000; **355:** 1412-8

Appendix D

Tissue Infectivity

When assessing whether an operation on a patient may contaminate surgical instruments with TSE consider whether the procedure has involved a tissue of high or medium infectivity.

Where one cannot be sure which form of TSE the patient is at risk of incubating assume that the risk is from variant CJD.

	Definite or Probable TSE	**Possible***	**At Risk of non vCJD#**	**At risk of vCJD#**
High risk Brain Spinal cord Posterior Eye	Destroy	Quarantine *	Destroy	Destroy
1. Medium risk Anterior eye Olfactory epithelium	Destroy	Quarantine *	Destroy	Destroy
2. Medium Risk Lymphoid Tissue**	Destroy	Quarantine *	Quarantine and contact IPCT	Destroy

*Quarantining may only occur after a detailed risk assessment from the ICPT/SDU. In general due to the logistical risks quarantining is not advocated.

**Lymphoid tissue is considered tonsil, appendix, lymph node, spleen, thymus, adrenal and rectum. Only variant CJD has not been identified in these tissues but for the sake of simplicity and clarity all TSEs will in general be treated in a similar manner unless a robust diagnosis of non-vCJD TSE has been achieved and the IPCT have risk assessed the situation.

#If in any doubt as to whether the risk is due to vCJD or other TSEs contact IPCT.

T

Appendix E

Sample questions to ask when eliciting whether an individual is in an at risk group for TSE

1. **Has anyone in your family ever suffered from Creutzfeldt-Jakob disease (CJD), GSS or Fatal Familial Insomnia (FII) or have you or anyone in your family previously had a genetic test showing them to be at increased risk of CJD, Gerstmann-Straussler-Scheinker (GSS) or FFI?**

2. **Have you ever had an operation on your brain or spine?**
   If so was it before September 1992? [If after August 1992 then the patient will not have received a contaminated dural graft. If the operation was earlier then the medical history will need to be considered in greater detail.]

3. **Have you ever been informed that you are at risk of developing CJD because of hormone injections that you received as a child (prior to 1986) or for fertility reasons?** (if female)
   Any recipients of contaminated hormone are likely to have been informed of their risk. If the individual has not been informed and has not received hormones in the past assume they are not at risk.

4. **Have you ever been informed that you have been exposed to CJD and may in certain circumstances e.g. operations pose a risk to others?**
   Enquire as to whether the exposure was to classical or variant CJD. If in doubt assume it to be variant CJD.
   In general individuals **at risk of incubating disease** will either know of their risk or it will be clear in their medical history.

Appendix F

Protocol for Management of Instruments and Tissues from Brain Biopsy Procedures

This appendix is a local modification to advice enclosed in a letter from the Chief Medical Officer dated 17th May 2004

Brain biopsy may reveal a diagnosis of TSE/CJD that was not suspected prior to the biopsy. This protocol minimises the risk that this could result in inadvertent exposure of subsequent patients to TSE agent. The consultant in charge of the case must take responsibility for coordination of the procedures and communications, including alerting the neuropathology laboratory in advance of sending samples and monitoring all aspects of the protocol.

Wherever possible avoid reusable instruments when biopsying the brain.

In the unlikely event they are needed follow the advice below regarding destruction or quarantining of instruments

A. Clinical assessment and biopsy procedure

1. This protocol should apply to all patients who have unexplained progressive dementia (or ataxia or neuropsychiatric syndromes) in whom diagnostic brain biopsy is considered appropriate in order to establish or exclude a diagnosis.
2. The neuro-radiology in such patients usually shows no evidence of a space-occupying lesion.
3. The patient does not fulfil the WHO criteria for probably or possible CJD. Indeed CJD may not have been considered on clinical grounds.
4. Brain biopsy, preferable an open block biopsy, is performed for diagnosis.

B. Handling the neurosurgical instruments

5. Single-use instruments should be used whenever possible without compromising patient safety.
6. Any reusable instruments that may have come into contact with brain or meninges should be washed at the point of use and quarantined immediately after use.

C. Handling the biopsy tissue

7. Inform neuropathology laboratory of imminent arrival of fresh high-risk tissue.
8. The tissue sample should be placed in a sterile empty biopsy container and double-bagged. Refer to “Labelling and transportation of samples which pose a potential infection Hazard”, Transfer to neuropathology laboratory, ideally within 10 minutes of collection.
9. In the neuropathology laboratory, using containment conditions appropriate for fresh human brain, remove a small portion of unfixed cortical grey matter

(at least 0.1g) from the biopsy. Store the sample frozen, clearly labelled and preferably at -70°C (-20°C is sufficient for storage for several weeks) in a designated freezer.

10. Fix the remainder of the tissue samples in formalin and treat with 96% formic acid for 1 hour post fixation (inactivation of infection risk) prior to processing into paraffin wax.

D. Neuropathological diagnosis and the fate of the Neurosurgical instruments

11. **If a definite diagnosis of a disorder other than CJD is made (**and there is no other evidence to suggest any form of CJD).

- The Neurosurgical instruments can be repossessed and reused.

12. **If a definite diagnosis of CJD is made**

- Destroy the Neurosurgical instruments and refer the case to NCJDSU by the clinician responsible for the patient. Precautions to minimise the risk of transmission as a result procedures carried out in the pathology laboratory and the clinic should be taken in accordance with the ACDP/SEAC TSE Guidance.

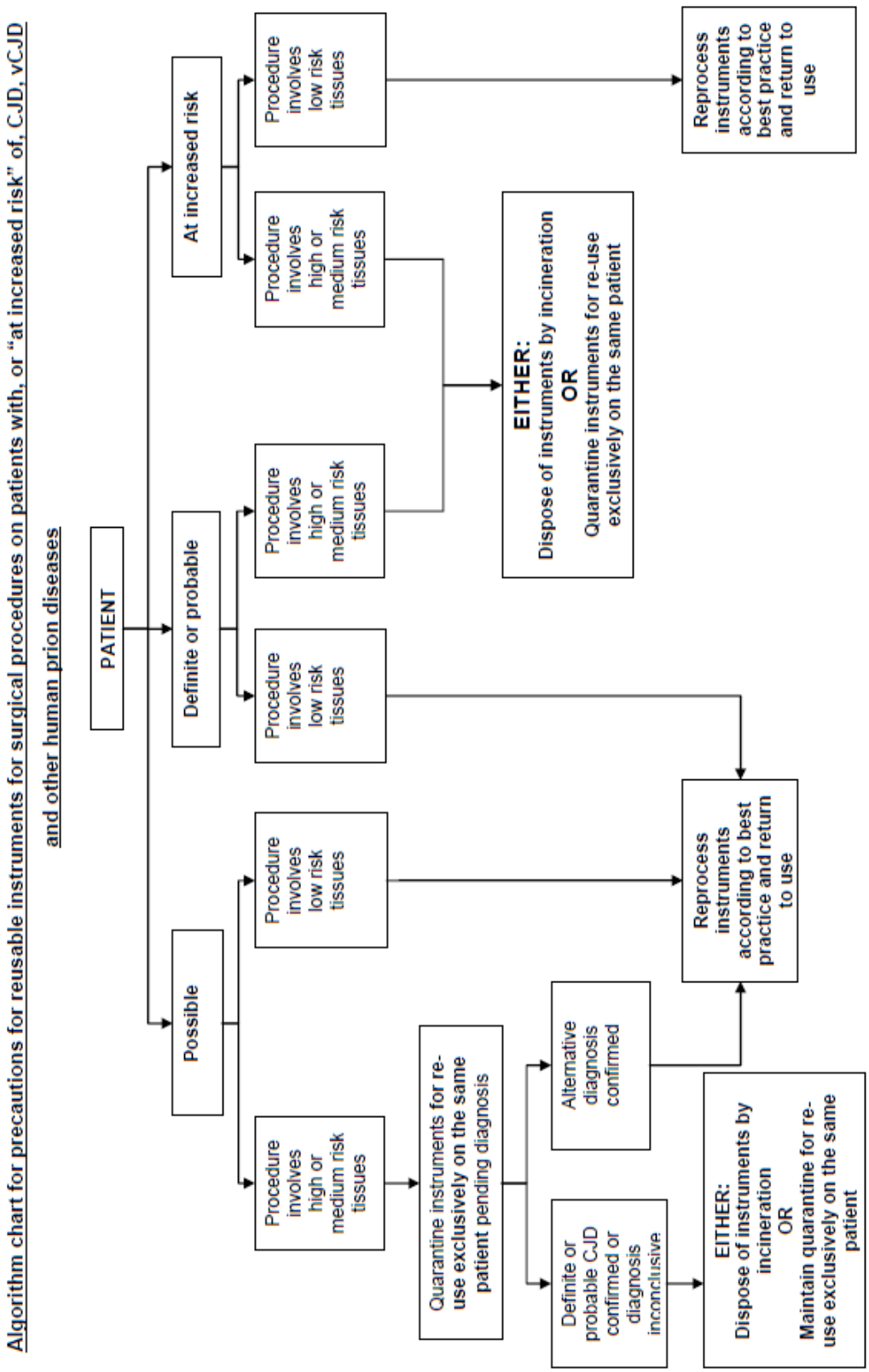
13. **If the diagnosis is uncertain**

- If the local neuropathologist cannot exclude CJD as a possible diagnosis, contact NCJDSU to refer the case for further investigation. NCJDSU will arrange uplift and transport of the fixed and frozen tissues.
- If an alternative diagnosis is made by the NCJDSU, the Neurosurgical instruments can be repossessed and reused.
- The neurosurgical instruments should be destroyed if a definite diagnosis of CJD is made by NCJDSU.
- If the diagnosis remains uncertain, the Neurosurgical instruments should remain in quarantine until a definite diagnosis is made, or the patient dies. Precautions to minimise the risk of transmission as a result of procedures in the pathology laboratory and the clinic should be taken in accordance with the ACDP/SEAC TSE Guidance.

14. If the patient dies without a diagnosis

- Seek consent for post mortem examination of the brain and, if it is given, follow the procedures set out in box 13 above.
- The instruments used for the biopsy should be destroyed, if consent for post mortem examination is not given.
- If the diagnosis is still uncertain after post mortem examination of the brain, the instruments used for biopsy should be destroyed.

Appendix G


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
PATIENT
At increased risk
Definite or probable
Possible
Procedure involves low risk tissues
Procedure involves high or medium risk tissues
Procedure involves low risk tissues
Procedure involves high or medium risk tissues
EITHER:
Dispose of instruments by incineration
OR
Quarantine instruments for re-use exclusively on the same patient
Reprocess instruments according to best practice and return to use
Procedure involves high or medium risk tissues
Quarantine instruments for re-use exclusively on the same patient pending diagnosis
Alternative diagnosis confirmed
Definite or probable CJD confirmed or diagnosis inconclusive
EITHER:
Dispose of instruments by incineration
OR
Maintain quarantine for re-use exclusively on the same patient
Procedure involves low risk tissues
Reprocess instruments according to best practice and return to use