St Helens and Knowsley Teaching Hospitals NHS
NHS Trust

INFECTION CONTROL MANUAL

Chapter 27

Creutzfeldt-Jakob Disease (CJD) and other Transmissible Spongiform Encephalopathies (TSE)

Recommending Committee:	**Hospital Control of Infection Committee**
Approving Committee:	**Clinical Performance Council**
Signature:	
Designation:	**Chairman Clinical Performance Council**
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INDEX

INTRODUCTION

Transmissible Spongiform Encephalopathies (TSEs), also known as prion diseases, are fatal degenerative brain diseases.

The name is derived as follows:

Transmissible: the disease can be transmitted from person to person
Spongiform: holes (vacuoles) appear in the brain, giving it a sponge-like appearance
Encephalopathy: abnormality of the brain

Examples of TSEs

In humans:	Creutzfeldt-Jakob Disease (CJD) New variant CJD (nvCJD) Kuru Gerstmann-Straussler-Scheinker syndrome (GSS) Fatal familial insomnia.
In cattle:	Bovine Spongiform Encephalopathy (BSE)
In sheep:	Scrapie

All human TSEs are very rare. The incidence of CJD is about 1 per million people each year. The other human TSEs are even more rare.

The agents responsible

TSEs are caused by unconventional infectious agents, currently thought to be infectious proteins, or prions, which do not have the same properties as bacteria or viruses.

Prions are concentrated in the brain, spinal cord and lymphoreticular tissues of the affected individual, causing dementia and death.
Prions have exceptional ability to withstand all conventional methods of decontamination e.g. heat and disinfectants.

Prions are NOT highly contagious and do not spread by normal contact. They spread by inoculation of infected neural tissue e.g. neurosurgery (on instruments or dura mater grafts), corneal transplant, injection of hormones (e.g. growth hormone/gonadotrophin) derived from the pituitaries of cadavers. There is no evidence of transmission to health careworkers during standard patient care.

Definitions of patient risk group

See Appendix A for diagnostic criteria and Appendix B for categorisation of patients by risk.

The prion protein is more widespread in the body of vCJD patients than patients with sporadic CJD. In sporadic CJD prion is restricted to the CNS. In vCJD prions may be found also in lymphoid tissues including tonsils, spleen, appendix, rectum, thymus and adrenal gland. Therefore precautions for surgical procedures differ.

Isolation precautions

The patient with CJD or other TSE can be nursed on the open ward. No special precautions are required for excreta, saliva or body secretions. Normal social or routine clinical contact does not present a risk to healthcare workers or other patients.

Clinical procedures

Phlebotomy/Drug administration by injection/Insertion of intravascular lines

It is not known whether CJD can be transmitted by blood. However, standard infection control procedures will minimise any risk. Take care during the handling and disposal of any sharps.
Phlebotomy: only experienced staff should take blood from these patients.
Drug administration by injection: only experienced staff should give injections to these patients.
Insertion of intravascular lines: only experienced staff should insert intravascular lines into these patients.

Wound re-dressing

Late stage CJD patients may suffer from pressure sores. These lesions should be dressed regularly, using standard infection control precautions. Contaminated dressings should be disposed of as clinical waste.

Lumbar puncture

Only experienced staff should carry out this procedure.
Wear gloves, plastic apron. Use eye protection if splashing may occur.
Use only single-use disposable instruments.
Affix Danger of Infection sticker to specimen(s) and request form(s) and inform the laboratory in advance.

Surgical procedure

The patient must be last on the operating list
Involve only the minimum number of staff required
Use disposable drapes and dressings

Protective clothing (in addition to standard operating theatre outfit)

Water repellent gown, over a plastic apron
Gloves
Mask and goggles, or full-face visor
Visor/goggles

All protective clothing must be destroyed by incineration after use.

Instruments

To determine whether the instruments need to be incinerated/quarantined/processed for re-use:

1. Check what category the patient falls into (Appendix A and B).
2. Determine tissue infectivity (Appendix C).
3. Then use either Appendix D (for CJD other than variant CJD) or Appendix E (vCJD) flowcharts.

Complex instruments

Expensive instruments may be prevented from being contaminated by using shield guards or coverings so that entire items do not need to be destroyed. Only parts in contact with high risk tissues and the protective coverings would need to be incinerated.

Laser techniques for tonsillectomy

There is no evidence of transmission of TSEs by the respiratory route. Any risk from smoke plumes is thought to be very low.

Childbirth

Involve only the minimum number of staff required
Use disposable drapes and dressings

Protective clothing (in addition to standard outfit)

Water repellent gown, over a plastic apron
Gloves
Mask and goggles or full-face visor
Visor/goggles

All protective clothing must be destroyed by incineration after use.

Instruments

Responsibilities: See HSDU/theatre operational procedures.
To determine whether the instruments need to be incinerated/quarantined/processed for re-use:

1. Check what category the patient falls into (Appendix A and B).

2. Determine tissue infectivity (Appendix C).
3. Then use either Appendix D (for CJD other than variant CJD) or Appendix E (vCJD) flowcharts.

Placenta

The placenta, other associated material and fluids should be treated as if infected and disposed of as infectious clinical waste by incineration, unless they are needed for investigation.

Linen: Used or fouled bed linen should be dealt with the same as for other patients. If sheets become contaminated with CSF they should be placed in yellow bags for incineration (double bag).

Waste disposal: Waste material should be handled the same as for waste from other patients (see Chapter 15, Infection Control Manual). Waste contaminated with high risk material e.g. CSF should be double bagged.

Spillages: Disposable gloves and plastic aprons must be worn. Use disinfectant appropriate to the type of spillage (see Disinfection Policy, Chapter 9, Infection Control Manual). Dilution is the most important element in cleaning up spillages.

Sharps injury: See Chapter 22, Infection Control Manual.

Specimens for the laboratory

Known/Suspected CJD

All specimens including biopsy material, urine faeces, swabs, blood and CSF must be labelled “Danger of Infection”. These precautions apply even to formalin fixed material, since formalin does NOT inactivate CJD.

At risk patients

Specimens eg, urine, faeces, swabs (but NOT CSF) from **at risk, asymptomatic patients** can be collected, processed and handled as for any other patient. CSF must be labelled “Danger of Infection”.

Visits to other departments

For non-invasive investigation eg X-ray, no specific precautions are required.

Reporting of suspected cases

From July 2004, a new national reporting system commenced. All new suspect cases of CJD and other prion diseases must be notified to the National Creutzfeldt Jakob Disease Surveillance Unit (NCJDSU) in Edinburgh and the National Prion Clinic (NPC) in London (see Contacts – Appendix F) with a copy to the local CCDC (see Appendix G

for form). Failure to notify would prevent early action to trace and withdraw blood donation and to institute other infection control measures.
Patients/families should be provided with a single information leaflet explaining the work of both the National CJD Surveillance Unit and the National Prion Clinic. (Appendix H)

On encountering patients whom they suspect to be suffering from CJD, or a related prion disease, consultant neurologists (or other clinicians) will:

- complete the national reporting form and obtain consent from the patient, lead relative or carer/patient representatives;
- fax or post the completed form, with consent, to the National CJD Surveillance Unit and the National Prion Clinic;
- advise the patient, carer, or independent representative that staff from the National CJD Surveillance Unit will visit the patient for national surveillance purposes (with their consent). They will also be given the opportunity to participate in research programmes operated by the Unit should they so wish;
- consider utilising the expertise in diagnosis and management of patients offered by staff at the National Prion Clinic;
- advise the patient, carer or independent representative that staff from the National Prion Clinic will visit and offer the opportunity to participate in therapeutic trials and/or other clinical research programmes, should they so wish.
- make available samples of blood and Cerebrospinal Fluid (CSF), and the results of Magnetic Resonance Imaging (MRI) scans to the two units.

Quarantining of surgical instruments
This applies to instruments used while awaiting confirmation of the diagnosis of CJD. Destroy single use instruments by incineration.

Re-usable instruments should be washed to removed gross soiling. Take care to avoid splashing and aerosols holding instruments under the surface of the water in a sink into which water is running and draining out continuously. Wear gloves and eye protection. Take care to avoid sharps injury.

Allow to air dry in disposable bag. Then seal in impervious rigid plastic container with close fitting lid (available from HSDU). Label with patients ID (Name, Hospital Number and DOB), the surgical procedure performed, and name of responsible person (e.g. theatre manager). Securely store in HSDU until the outcome of any investigation is known. If confirmed CJD, incinerate without further examination.

Only if an alternative definitive diagnosis is confirmed, remove from box by responsible person, decontaminate in usual way and return to normal use.

Keep record of all action taken.

Staff records

Occupational Health Records will need to record employees exposed to CJD (not routine clinical exposure).

Examples:

Staff performing invasive clinical procedures on patients suspected to be suffering CJD, especially where there is risk of exposure to CNS/eye.

Laboratory staff handling tissue specimens (not blood).

Staff undertaking postmortem examinations.

The list must include

- Name (full name and maiden name), NI number, DOB, dates of employment
- Type of work
- Any specific exposure/accident/incident which may be reportable under RIDDOR

The list must be kept for 40 years. Staff must report any such exposures to Occupational Health.

After death

A body bag is required (see Chapter 16 Infection Control Manual). The relatives may view the body. After post mortem has taken place viewing and possible superficial contact such as touching or kissing need not be discouraged. Body bags may be rolled down temporarily to allow superficial contact. There is no need to deny relatives this opportunity if a post mortem has been performed. None of the patient's tissues may be used for transplant purposes. Embalming should be avoided. There is no need for special arrangements for burial or cremation.

References

Transmissible Spongiform Encephalopathy agents: safe working and the prevention of infection.
Advisory Committee on Dangerous Pathogens Spongiform Encephalopathy Advisory Committee, 1998.

Revised TSE Guidance (June 2003).

APPENDIX A

Classification criteria

Sporadic CJD

Definite: Neuropathological/immunocytochemical confirmation is required for a diagnosis of definite sporadic CJD.

Probable: 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).

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

Iatrogenic CJD

Iatrogenic CJD patients display progressive cerebellar syndrome in a pituitary hormone recipient **or** sporadic CJD with a recognised exposure risk (eg.dura mater transplant). A definite diagnosis or iatrogenic CJD still require a neuropathological examination.

Familial CJD

Patients with familial CJD will have definite or probable CJD (as 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)

Definite: 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 cerebellum.

Probable: 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 symmetrical high signal in the posterior thalamus on a MRI brain scan.

   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. They will **also** have at least four out of five of the symptoms listed in (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 fulfil the criteria for possible CJD

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 it 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.

APPENDIX B

Categorisation of patients by risk

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

1. Symptomatic patients	1.1 Patients who fulfil the diagnostic criteria for definite, probable or possible CJD or vCJD (see Appendix A 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 genetic mutations	2.1Individuals who have or have had two or more blood relatives affected by CJD or other prion disease, or a relative known to have a genetic mutation indicative of familial CJD. 2.2 Individuals who have been shown by specific genetic testing to be at significant risk of developing CJD or other prion disease.
3. Asymptomatic patients potentially at risk from iatrogenic exposure.	3.1 Recipients of hormone derived from human pituitary glands e.g. growth hormone, gonadotrophin 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 because of exposure to instruments used on, or receipt of blood, plasma derivatives, organs or tissues donated by, a patient who went on to develop CJD or vCJD.

APPENDX C

Categorisation of tissue infectivity

CJD other than vCJD

Tissue infectivity	Status of patient			
	Definite/probable	Possible	At risk	
			Genetic	Iatrogenic
High: ❑ Brain ❑ Spinal cord ❑ Posterior eye	**D**	**Q**	**D**	**D**
Medium: ❑ Anterior eye ❑ Olfactory epithelium	**D**	**Q**	**D**	**D**
Low/none detectable	**NSP**	**NSP**	**NSP**	**NSP**

vCJD

Tissue infectivity	Status of patient			
	Definite/probable	Possible	At risk	
				Iatrogenic
High: ❑ Brain ❑ Spinal cord ❑ Posterior eye	**D**	**Q**		**D**
Medium: ❑ Lymphoid tissue ❑ Anterior eye ❑ Olfactory epithelium	**D**	**Q**		**D**
Low/none detectable	**NSP**	**NSP**		**NSP**

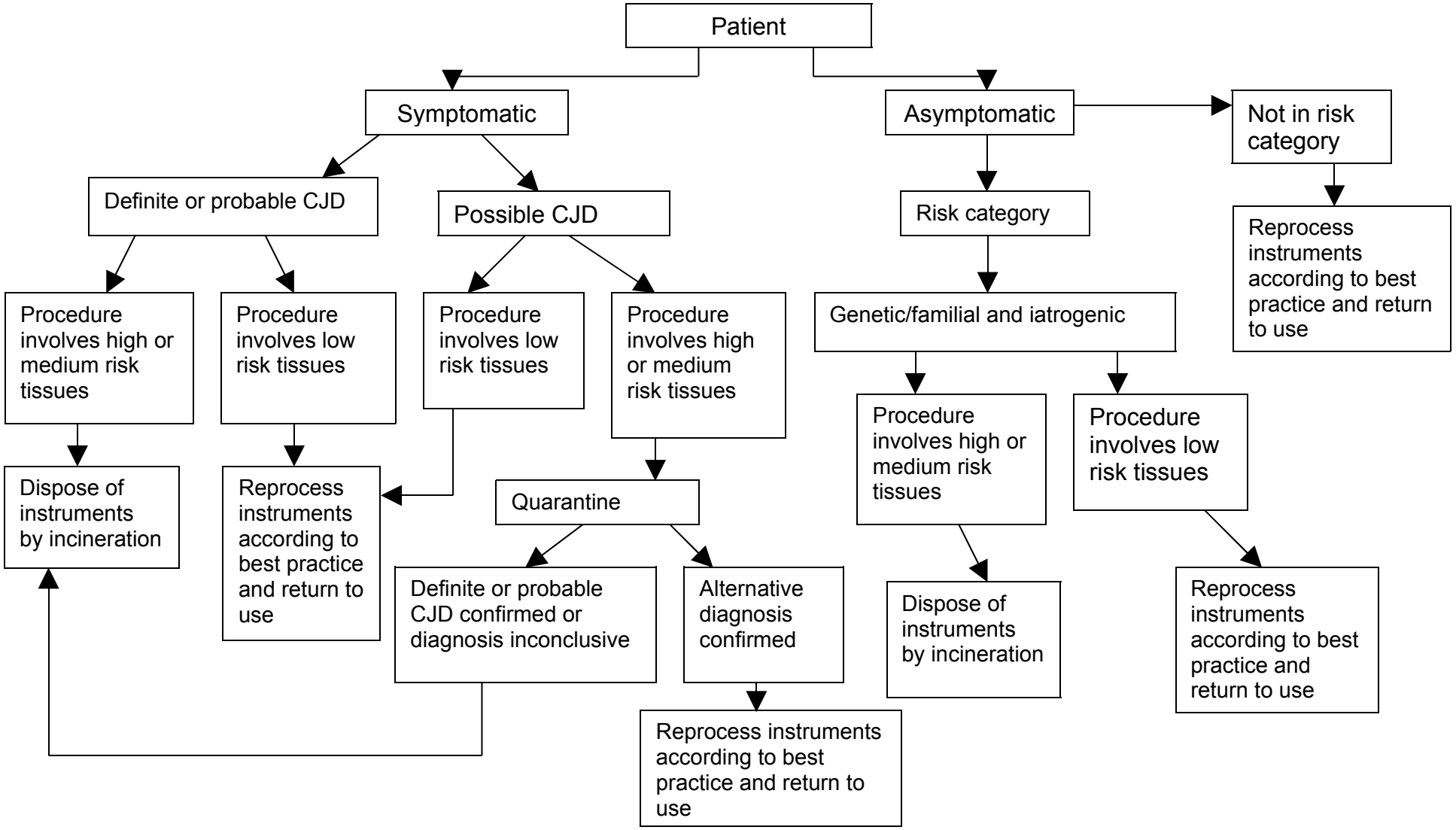
***** PrP-res has been detected in tonsil, appendix, lymph node, spleen, thymus, adrenal gland and rectum of vCJD patients.

Key

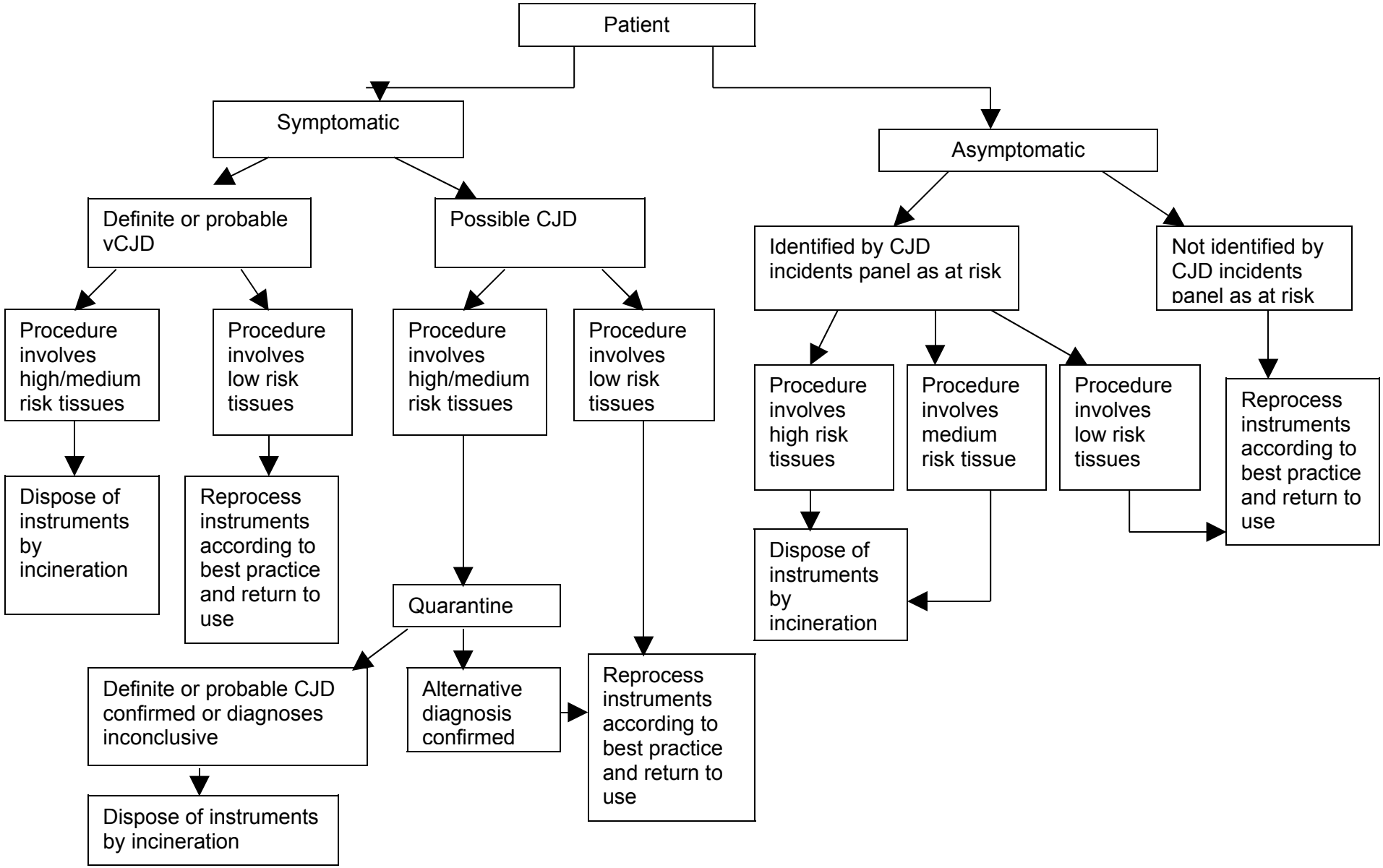
D = destroy by incineration
Q = quarantine pending diagnosis
NSP= no special precautions

APPENDIX D

ALGORITHM CHART FOR PRECAUTIONS FOR SURGICAL PROCEDURES ON KNOWN, SUSPECT OR AT RISK PATIENTS:
CJD OTHER THAN VARIANT CJD



ALGORITHM CHART FOR PRECAUTIONS FOR SURGICAL PROCEDURES ON KNOWN, SUSPECT OR AT RISK PATIENTS: VARIANT CJD



USEFUL CONTACTS APPENDIX F

- The National CJD Surveillance Unit in Edinburgh can provide advice on clinical and neuropathological aspects of CJD.It should be notified of any patient suspected on clinical grounds of having CJD. It can be contacted at::

  Director, National CJD Surveillance Unit
  Western General Hospital
  Crewe Road
  Edinburgh EH4 2XUT
  Tel: 0131 537 2128 (Clinical office)
  0131 537 1980 (Pathology Telephone)
  Fax: 0131 343 1404
  Email: jan.mackenzie@ed.ac.uk

- The National Prion Clinic at St Mary's Hospital, London specialises in the care of patients suffering from CJD. It should be notified of any patient suspected on clinical grounds of having CJD. It can be contacted at:

  National Prion Clinic
  Department of Neurology
  St Mary's Hospital
  Praed Street
  London W2 1NY
  Tel: 020 7886 6883
  Fax: 020 7061 9889
  Email: help.prion@st-marys.nhs.uk

- The CJD Support Network is a voluntary organisation set up to provide help and support for patients of all types of CJD and their families. The Network had undertaken a case co-ordination initiative aimed at facilitating the co-ordination enabling cost effect care and ensuring appropriate responses to carer's needs. It can be contacted at:

  National CJD Co-ordinator
  CJD Support Network
  PO Box 346
  Market Drayton, Shropshire TF9 4AR
  Tel: 01630 673 993

- The Human BSE Foundation is a voluntary organisation run by families of vCJD patients aimed at helping relatives, friends and carers of vCJD patients by providing support, information and practical advice. It can be contacted at:

The Human BSE Foundation
Matfen Court,
Chester Le Street, County Durham, DH2 2TX
Tel (helpline):0191 389 4157
www.hbsef.org

NATIONAL CJD REPORTING FORM **APPENDIX G**

Fax to: NCJDSU 0131 343 1404, NPC 0207 061 9889 and also to your local CCDC when completed.

Patient details
Surname:……Forename(s):……
Postal address:……
……
……
……
Postcode:……...

Telephone number:……..

Fax Number:…….

Email Address:……...

NHS Number, if known:……
……

Family, carer or independent representative details (if appropriate*)

* This may be appropriate if the approach is made via a lead family member, carer or independent representative (i.e. when a patient is too ill to be approached directly or has a preference for this route).
Surname:…….Forename(s):……
Postal Address:……
…
……
……
Postcode:……...

Telephone number:……..

Fax Number:…….

Email Address:……...

Neurologist details (or other hospital clinician) Surname:…… …….Forename(s):……..

Hospital Postal Address:…… …….. …… ……

Postcode:……..

Telephone number:……..

Fax Number:…….

CCDC details

Surname:…….Forename(s):……

Postal Address:…….. .. ……

Postcode:……...

Telephone number:……..

GP Details

Surname:…….Forename(s):……

GP Practice Postal Address:…… ……... …… ……

Postcode:……...

Telephone number:……..

Fax Number:…….

Brief clinical details: (please attach recent letter or discharge summary)

……
……
……
……

Consent:

*please delete as appropriate

I have been provided with the patient information leaflet which explains the roles of the National CJD Surveillance Unit and the National Prion Clinic.

YES/NO

I agree to my/the patient's* details being forwarded to the National CJD Surveillance Unit and the National Prion Clinic.

YES/NO

I agree that staff from the National CJD Surveillance Unit in Edinburgh and the National Prion Clinic in London can visit myself/the patient* and my/their* relatives at a mutually convenient time for clinical assessment and surveillance purposes and to provide the opportunity, should we wish, to discuss ongoing research, including clinical trials of potential treatments.

YES/NO

I understand that this may mean providing further information to help in the organisation of my/the patient's* care, and to contribute to a better understanding of the illness.

YES/NO

Signed:……

Print:……

Date:……

On completion, please fax to NCJDSU 0131 343 1404, NPC 0207 061 9889 and also to your local CCDC.

PATIENT/CARER INFORMATION SHEET **APPENDIX H**

WHY HAVE I RECEIVED THIS INFORMATION?

You are being given this information because the possibility of CJD or another form of prion disease is being considered. Two major, publicly funded, specialist teams work in the UK on these diseases. With your consent, someone from each team would like to visit you/the patient and discuss how they can help and also to discuss important research they are doing into the causes of, and treatments for, prion diseases.

Some people referred to these teams will turn out not to have prion disease. Whatever diagnosis is confirmed, your/the patients' care will be lead by your local clinical service. Involvement in any research is entirely voluntary and refusal to participate will not affect your care in any way.

WHO WILL BE APPROACHING ME?

11) The National CJD Surveillance Unit team

Western General Hospital, Edinburgh
Tel: 0131 537 2128
http://www.cjd.ed.ac.uk/

The National CJD Surveillance Unit (NCJDSU) is funded by the Department of Health to identify, classify and investigate prion diseases in the United Kingdom. The NCJDSU team will want to visit to make sure that it is a prion disease and also to identify what type of prion disease may be affecting you/the patient. They will also wish to discuss other research projects. The team consists of a doctor and a nurse who will travel to see you/the patient and to discuss things with the local clinicians. They can provide advice, guidance and diagnostic tests to help the doctor looking after you/the patient both in making the diagnosis and in giving care. A National Care Team is based at the NCJDSU. If you and the local clinicians wish, a care co-ordinator from the National Care Team will visit later to assess your/the patient's care needs and to help you and the local doctors to identify ways to meet these needs if they are not already met.

1

22) The National Prion Clinic and MRC Prion Unit treatment team

National Hospital for Neurology and Neurosurgery, London
Tel: 020 7886 6883
http://www.st-marys.nhs.uk/specialist/prion/index_prion.htm

The National Prion Clinic offers specialist in- and out-patient services including diagnostic facilities which are available to help support your doctors, if they wish, in choosing the best type of care for you as well as providing specialised advice and counselling.

A major, long-term, research effort is underway at the MRC Prion Unit in London to find new treatments for prion disease and to study if these treatments benefit patients (the MRC Prion-1 trial). We need to understand better how these rare diseases progress in everyone, not just those taking a new drug(s), in order to make real progress towards an effective treatment. Therefore, the Prion-1 trial has been designed to allow all patients that want to participate to do so, irrespective of whether or not they want to try the drug

being tested at that time, or what form of prion disease they might have. The Department of Health has made a major financial investment in the trial and asked the

Medical Research Council, which has over 50 years experience running trials, to find the best treatments for these diseases. The team would like to visit you to discuss the treatment trial and other research projects aimed at tackling these diseases.

IS THERE ANYONE ELSE I CAN TALK TO?

Many patients and carers find it helpful to talk to others with experience of the same diseases. There are two UK support groups for patients with prion disease and their families and carers. Both have confidential helplines, offering support and practical information. They each have a website, and regular newsletters, with lots of useful information.

Because these diseases are very rare, many local medical and care services have no experience of them. But you are not alone. The support groups, together with the specialist centres, are committed to making sure you don't miss out on whatever help is available. You don't have to wait until you have a firm diagnosis - support is there, if you want it, from the moment you have to consider the possibility of prion disease.

The CJD Support Network is a UK charity for people with any type of CJD.
Helpline: 01630 673973
http://www.cjdsupport.net/index.html

The Human BSE Foundation is a UK charity primarily for those who have (or may have) contracted variant CJD.
Helpline: 0191 389 4157
http://www.hbsef.org/