Protocol for Reporting and Management of cases of **Creutzfeldt Jakob Disease (CJD)** and other **Transmissible Spongiform Encephalopathies (TSEs)** or of a person at increased risk of a TSE

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Protocol for Reporting and Management of cases of Creutzfeldt Jakob Disease (CJD) and other Transmissible Spongiform Encephalopathies (TSEs) or of a person at increased risk of a TSE[1]: 2019

This protocol guides on the follow up required following a report of:
- A newly diagnosed or suspected case of CJD/vCJD
- A person at increased risk of CJD/vCJD[2]
- An invasive procedure carried out on a patient with CJD/vCJD or at increased risk of CJD/vCJD where TSE infection control guidelines were not followed

In December 2018, the Infectious Diseases (Amendment) Regulations 2018 was signed into law. The Regulations amended the Infectious Diseases Regulations 1981 to require both confirmed or suspected cases of Creutzfeld Jakob disease and variant Creutzfeld Jakob disease to be notifiable (reported to the Medical Officer of Health).

Cases classified as "Diagnosis unclear" (Section 11) must also be investigated unless advised otherwise by the Incident Panel on Transmissible Spongiform Encephalopathies (IPTSE)/neurologist on the IPTSE.[3]

The key aims of actions taken are to:
- Prevent secondary transmissions of TSE
- Provide appropriate diagnostic procedures for the patient
- Determine if a case is due to iatrogenic transmission
- Protect confidentiality of patient information

Thus key actions recommended are:
- Prompt infection prevention and control procedures
- Appropriate diagnostic procedures
- Prompt involvement of the National CJD Surveillance Unit (NCJDSU), Beaumont Hospital, Dublin for referral of cerebrospinal fluid sample and confirmatory post-mortem diagnosis
- Prompt reporting by treating clinician to the Medical Officer of Health (MOH) in the local Department of Public Health[4] and to the Incident Panel on Transmissible Spongiform Encephalopathies (IPTSE).

Please note that the following process steps may not necessarily occur in chronological order. Also some duplication of reporting will be noted within the guidance- this is in order to ensure necessary communications.

Please note that:
1. The abbreviations CJD and vCJD will be used within this document for ease of readability. The specific types of TSE will be addressed where relevant.

   In this scenario CJD represents: iatrogenic (excludes iatrogenically acquired vCJD), genetic, sporadic, Fatal Familial Insomnia and Gerstmann–Sträussler–Scheinker syndrome. vCJD represents variant CJD and includes iatrogenically acquired vCJD.

2. The abbreviation MOH (Medical Officer for Health) will be used throughout this document to represent either the Director of Public Health (DPH) or a Consultant in Public Health Medicine (CPHM).

[1] Main reference for this document is Public Health Action Following a Report of a New Case of CJD or a Person at Increased Risk of CJD https://assets.publishing.service.gov.uk/government/uploads/system/uploads/attachment_data/file/474338/CJD_public_health_action_new_case_301015.pdf

[2] See Section 1

[3] Neurologist and member of the Incident Panel on Transmissible Spongiform Encephalopathies (IPTSE): See Appendix 10 for contact details.

[4] Notifications should be made to the Medical Officer for Health (MOH) in the local Department of Public Health. See Appendix 10 for contact details. The MOH, which may be the Director of Public Health (DPH) or Consultant in Public Health Medicine (CPHM), will inform/refer to the MOH in the area of residence of the case. Practicalities of case management will guide which department takes the lead.

*Please note the need for investigative teams to review an incident in line with HSE Safety Incident Management Policy 2014 https://www.hse.ie/eng/about/qavd/incident-management/

What is a CJD/vCJD incident?
Incidents involving the potential transmission of a TSE between patients through invasive clinical procedures, including surgery, endoscopy, blood donations, and organ and tissue donations.

A CJD or vCJD incident may occur when:

- A patient has donated organs/tissues before being diagnosed with CJD or vCJD
- A patient has donated blood before being diagnosed with vCJD
- A patient has donated organs/tissues before being identified as having an increased risk of CJD or vCJD[6]
- A patient has donated blood before being identified as having an increased risk of vCJD
- A patient with confirmed/probable/possible diagnosis of CJD or vCJD has had an invasive procedure involving high or medium level risk tissues within the likely infective period and appropriate infection control guidance was not followed
- A patient with an increased risk of CJD or vCJD had an invasive procedure involving high or medium level risk tissues and appropriate infection control guidance was not followed

Table A: Level of tissue infectivity by CJD type

Level of tissue infectivity by CJD type	Tissues
High infectivity for all CJD types	Brain, spinal cord, cranial nerves (entire optic nerve, intracranial components of other cranial nerves), cranial nerve ganglia, posterior eye (hyaloid face, retina, retinal pigment epithelium choroid, subretinal fluid, optic nerve), pituitary gland
Medium infectivity for all CJD types	Spinal ganglia and olfactory epithelium
Medium infectivity for Variant CJD and type uncertain (and vCJD is being considered)	Tonsil, appendix, spleen, thymus, adrenal gland, lymph nodes, gut associated lymphoid tissues (including the rectum)
Low infectivity for all CJD types	All other tissues not listed above are considered to have low levels of infectivity for all types of CJD

General recommendation
Where feasible preferred use of single-use devices is advised, for example disposable nasendoscopy scopes.

[6] See Section 1

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LIST OF ABBREVIATIONS

14-3-3 test	14-3-3 protein CSF immunoassay
CDU	Central Decontamination Unit
CIT	Case Investigation Team
CJD	Creutzfeldt-Jakob Disease
CPHM	Consultant in Public Health Medicine
CSF	Cerebral spinal fluid
DCDU	Dental Central Decontamination Unit
EDU	Endoscope Reprocessing Unit
FFI	Fatal Familial Insomnia
fCJD	Familial / genetic Creutzfeldt-Jakob Disease
GP	General Practitioner
GSS	Gerstmann–Sträussler–Scheinker syndrome
HIT	Hospital Investigation Team
HPSC	Health Protection Surveillance Centre
IBTS	Irish Blood Transfusion Service
iCJD	Iatrogenic Creutzfeldt-Jakob Disease
IPTSE	Incident Panel on Transmissible Spongiform Encephalopathies
LDU-D	Local Decontamination Unit Dental
MOH	Medical Officer of Health
NCJDSU	National CJD Surveillance Unit
PM	Post Mortem
RIMD	Reusable Invasive Medical Devices including flexible endoscopes
RT QuIC test	Real-time quaking-induced conversion CSF test
sCJD	Sporadic Creutzfeldt-Jakob Disease
TSE	Transmissible spongiform encephalopathies
UK	United Kingdom
vCJD	Variant Creutzfeldt-Jakob Disease

USEFUL RESOURCES:

Endoscopes:
Management and decontamination of flexible endoscopes (HTM 01-06)
https://www.gov.uk/government/uploads/system/uploads/attachment_data/file/530418/HTM0106_PartA.pdf

Annex F Endoscopy
https://assets.publishing.service.gov.uk/government/uploads/system/uploads/attachment_data/file/470292/ACDP_TSE_Annex_F_Oct_2015.pdf

Surgical Instruments:
Health Technical Memorandum (HTM) 01-01: management and decontamination of surgical instruments (medical devices) used in acute care. https://www.gov.uk/government/publications/management-and-decontamination-of-surgical-instruments-used-in-acute-care

Annex M Managing vCJD risk in general surgery and liver transplantation
https://assets.publishing.service.gov.uk/government/uploads/system/uploads/attachment_data/file/209780/Annex_M_-_Mananging_vCJD_risk.pdf

Annex E Quarantining of surgical instruments https://assets.publishing.service.gov.uk/government/uploads/system/uploads/attachment_data/file/547149/Annex_E_August_2016.pdf

General Guidance:
Annex C General principles of decontamination and waste disposal
https://assets.publishing.service.gov.uk/government/uploads/system/uploads/attachment_data/file/427855/Annex_C_v3.0.pdf

Part 4 Infection Control and Prevention of CJD and VCJD in Healthcare and community Settings
https://assets.publishing.service.gov.uk/government/uploads/system/uploads/attachment_data/file/427854/Infection_controlv3.0.pdf

Public Health Action Following a Report of a New Case of CJD or a Person at Increased Risk of CJD
https://assets.publishing.service.gov.uk/government/uploads/system/uploads/attachment_data/file/474338/CJD_public_health_action_new_case_301015.pdf

Please refer to https://www.gov.uk/government/organisations/public-health-england to check for updated guidance documents

HSE:
Health Protection Surveillance Centre
http://www.hpsc.ie/a-z/other/cjd/

HSE Safety Incident Management Policy 2014
https://www.hse.ie/eng/about/qavd/incident-management/safety-incident-management-policy-2014-with-addendum-jan-2017.pdf

Decontamination of reusable invasive medical devices
https://www.hse.ie/eng/about/who/qid/quality-and-patient-safety-documents/deccont-rimd.html

TRANSMISSIBLE SPONGIFORM ENCEPHALOPATHIES

Transmissible spongiform encephalopathies (TSEs) are fatal degenerative brain diseases that occur in humans and some animal species. The causative agent is a protease resistant protein, which is an altered form of naturally occurring prion protein (PrP). PrP is normally present in human and animal brain tissue. In TSEs the altered form (also known as the scrapie agent or PrPSc) accumulates in the brain. These altered prion proteins are remarkably resistant to inactivation by standard chemical, thermal and other means of inactivating microorganisms. Prion diseases lead to symptoms of brain dysfunction, including difficulties with movements, memory problems and dementia. Currently, there are no treatments that have been shown to halt progression or to reverse the disease.

Five human forms of TSE have been reported to date:

- **Sporadic Creutzfeldt–Jakob disease (sCJD)** was first described in the 1920's, and is the most common form of human TSE but is still rare. It has a worldwide incidence of about 1 case per million per year. In the Republic of Ireland there is a notified range of 1-7 cases per year.[7]
- **Variant CJD (vCJD)** was identified in March 1996 by the UK National CJD Research and Surveillance Unit in Edinburgh. This form of CJD differed from previously recognised types as the patients affected were younger, their symptoms were different and the appearance of their brain tissue after death was not the same as in the sporadic form. Research findings published since March 1996 indicate that vCJD and bovine spongiform encephalopathy (BSE) in cattle are caused by the same infectious agent.
- **Iatrogenic Variant CJD** (the transmission of variant CJD through the use of past medical treatments) is reported to have occurred via transfusion of non-leucodepleted red blood cells in the 1990s (4 instances) and UK plasma used to produce Factor VIII (1 instance).[8]
- **Iatrogenic CJD (iCJD)** result from transmission during past medical interventions and include examples such as Dura mater grafts (~110 cases worldwide), Human Growth Hormone (~130 cases worldwide) and Neurosurgery contaminated medical equipment (~7 cases worldwide).
- **Genetic TSE** (also called inherited or familial prion diseases) are of autosomal dominant inheritance, account for only 10% of all TSE cases, and are linked to mutations in the prion protein coding sequence (*PRNP*). Mutations in the PrP gene (PRNP) makes the conversion into the abnormal form of the protein (PrPSc) more likely. Gerstmann-Sträussler-Scheinker syndrome (GSS) and Fatal Familial Insomnia (FFI) are types of Genetic TSE caused by mutations at different locations on the *PRNP* gene. GSS patients suffer predominantly from problems of balance and incoordination, affecting ~1 person in 10 million per year while FFI is characterised by abnormal sleeping patterns and is much rarer. For a list of all currently known *PRNP* mutations associated with prion disease please see the Diagnostic Criteria.
- **Kuru** occurred at epidemic levels during the 1950s-60s among the Fore people in the highlands of Papua New Guinea. The disease was the result of the practice of ritualistic cannibalism among the Fore, in which relatives prepared and handled the tissues (including brain) of deceased family members.

Number of CJD notifications from December 1996 to 2016*

	Number of notifications from December 1996 to 2016
Sporadic	76 (1-7 per year)
Iatrogenic	2
Familial	2
vCJD	4 (last case notified 2006)

* Data presented are based on notifications from HSE Areas and from the Irish National Creutzfeldt-Jakob Disease Surveillance Unit. Annual figures here are based on the year the notification was entered on the Computerised Infectious Disease Reporting (CIDR) system and consequently may differ from annual figures published by the Irish National Creutzfeldt-Jakob Disease Surveillance Unit.

7 http://www.hpsc.ie/a-z/other/cjd/publications/annualreports/

8 Source: https://webarchive.nationalarchives.gov.uk/20140714111136/http://www.hpa.org.uk/webc/HPAwebFile/HPAweb_C/1274091065209
"Infection with vCJD has probably been transmitted to four patients through blood transfusions in the UK from three donors who were diagnosed with vCJD after donating the blood. One of these patients had not developed clinical disease before dying from another cause. All four cases had received transfusions of non-leucodepleted red blood cells between 1996 and 1999. There have been no new cases of vCJD linked to blood transfusion since 2006.
Sporadic CJD has been monitored for many years in many countries. To date, no other forms of human prion disease, including sporadic CJD, have been transmitted by blood transfusions".

Please note that the abbreviation CJD and vCJD will be used within this document for ease of readability. The specific types of TSE will be addressed where relevant.

In this scenario CJD includes iatrogenic (excludes iatrogenically acquired vCJD), genetic and sporadic. vCJD includes iatrogenically acquired vCJD.

Risk assessments have considered evidence on tissue infectivity and the effectiveness of routine decontamination for prion protein removal and have concluded that the risks of CJD and vCJD transmission via surgery appear significant. **These conclusions and the lack of a reliable method to identify CJD or vCJD infection during the asymptomatic period continue to support a precautionary approach to the management of potential exposures.**[9]

Cerebrospinal fluid (CSF) tests for the investigation of patients with suspected sporadic CJD:

The CSF laboratory of the National CJD Research & Surveillance Unit (NCJDRSU Edinburgh) provides ROI with a diagnostic service for the analysis of CSF.

They perform two tests on the CSF:

1. 14-3-3 protein test. The current sensitivity and specificity for CSF 14-3-3 is 86% and 94% (NCJDRSU, Edin).

2. Real-time quaking induced conversion (RT-QuIC). This newer test is being done on all samples since January 2019. This is a very sensitive (92%) and specific (99%) test for the diagnosis of sporadic Creutzfeldt-Jakob disease.

This test is now included as part of the diagnostic criteria.

Please note samples are only to be sent through Beaumont Laboratory or Cork University Hospital.

9 *Public Health Action Following a Report of a New Case of CJD or a Person at Increased Risk of CJD* https://assets.publishing.service.gov.uk/government/uploads/system/uploads/attachment_data/file/474338/CJD_public_health_action_new_case_301015.pdf

SECTION 1: AT INCREASED RISK OF CJD/VCJD

Patients identified to be at increased risk include*:

Related to blood transfusions

- People who have received blood or blood components from someone who went on to develop vCJD
- People who have given blood or blood components to someone who went on to develop vCJD
- People who have received blood or blood components from someone who has also given blood or blood components to a patient who went to develop vCJD
- People who have received blood or blood components from 300 or more UK donors since 1990.

Related to surgery

- People who have had surgery using instruments that had been used on high or medium level risk tissues of someone who developed CJD
- People who underwent an intradural neurosurgical or intradural spinal procedure **before July 1996 who received (or might have received) a graft of human-derived dura mater**.
- People who have received an organ or tissue from a donor infected with CJD or at increased risk of CJD

Related to other medical care

- **Treatment with certain UK sourced plasma products between 1990 and 2001 (inclusive)**
- People who have been treated with **growth hormone from UK sourced human pituitary glands (before 1987)**
- People who have been treated with **gonadotrophin sourced from human pituitary glands for fertility treatment (before 1987)**
- People who have been told by a specialist that they have a risk of developing the genetic form of CJD

Patients identified not to be at increased risk of CJD:

Blood relatives who have been tested and **do not have** a disease specific mutation in the prion protein gene are not at increased risk of CJD. Incidents involving these patients should not be reported.

*Adapted from the UK reference document which is found at:
https://www.gov.uk/government/uploads/system/uploads/attachment_data/file/271414/Frequently_asked_questions.pdf

SECTION 2: PRE-OPERATIVE ASSESSMENT

The following patients should have their CJD/vCJD risk assessed and documented as part of the routine pre-operative assessment process:

All patients about to undergo:

1. **a neurosurgical procedure (including spinal and neuro-endoscopy procedures)**

or

2. **an ophthalmological procedure likely to involve contact with tissues of potentially high levels of TSE infectivity,**

Tissues of potentially high levels of TSE infectivity are**:**

Brain

Spinal cord

Dura mater

Cranial nerves, specifically:
- the entire optic nerve
- the intracranial components of the other cranial nerves

Cranial nerve ganglia

Posterior eye, specifically:
- posterior hyaloid face
- retina
- retinal pigment epithelium
- choroid
- subretinal fluid
- optic nerve

Pituitary gland

A draft pre-operative risk assessment form can be found in Appendix 1. Many hospitals will already be utilising their own version.

The purpose of the pre-operative risk assessment is to:
- Determine if the case has **known risk factors for** developing CJD or vCJD such as per Section 1.
- Determine if the case is diagnosed with/suspected to have CJD or vCJD.

Further information can be found in Appendix 2 and please see algorithm in Appendix 6.

Appendix 3 is a draft information sheet for patients on this preoperative assessment for CJD/vCJD risk. This is optional and for use if required.

NB:
If a patient is thought to be at increased risk for a TSE, further investigation should be done.

If following this local investigation, the increased risk is considered still possible, this assessment needs to be discussed in detail with the IPTSE.

If the patients procedure is routine and can be delayed to allow consultation with the IPTSE, that should occur. If procedure cannot be delayed special infection prevention and control precautions should be taken for the patient's procedure including quarantining of instruments, and the local infection prevention and control team should be consulted for advice.

Only when this full risk assessment is done in conjunction with the IPTSE, should the patient be informed. Please note that certain healthcare institutions whose patients are in specific groups at increased risk of a TSE may utilise a wider range of pre-operative assessments.

SECTION 3: ROLE OF THE TREATING CLINICIAN

3.1 Where a treating clinician considers CJD or vCJD in a patient's differential diagnosis:

All efforts should be made by the treating clinician to promptly ascertain the likelihood of a CJD diagnosis and ideally before any procedures are carried out.

- An experienced neurologist should be directly involved in the case assessment.
- Review of all data is required including clinial information, CSF, EEG and MRI.
- Where uncertainty remains the treating clinician should liaise as soon as possible with the IPTSE neurologist. The case may require discussion with the IPTSE.

3.2 Where a treating clinician considers CJD or vCJD in a patient's differential diagnosis or receives confirmation of a diagnosis of CJD or vCJD, complete Form A (Appendix 11) and send a copy to the following:

- NCJDSU (Form A can also be used as the referral form for 14-3-3 CSF testing by the NCJDSU)
- MOH (local Department of Public Health)
- IPTSE

Contact details can be found in Appendix 10.

Notes on completing Form A

- Completion and forwarding of Form A is essential and should be done as soon as CJD or vCJD is considered in a patient's differential diagnosis (ideally within one working day)
- If case is symptomatic, classify the patient according to the Diagnostic Criteria (Appendix 4 for guidance)
- Both confirmed or suspected cases of CJD and vCJD are notifiable (reported to the medical officer of health).
- Prompt reporting to the MOH is important to enable prompt infection prevention and control procedures
- If the patient meets the criteria for "Diagnosis unclear" (i.e. the diagnostic criteria for definite, probable or possible CJD or vCJD are not met, nor is there a reasonable alternative diagnosis, however, CJD remains a possibility), please refer to Section 11.

3.3 Determine if the case has known risk factors for developing CJD or vCJD (refer to Section 1).

3.4 Contact the hospital's CEO to establish a Hospital Investigation Team (HIT).* This team will initially determine if an incident may have occurred."

3.5 Where the case has died:

- Report the case to the coroner
- Inform the NCJDSU
- Inform the local Department of Public Health

*Please note the need for investigative teams to review an incident in line with HSE Safety Incident Management Policy 2014.

SECTION 4: ROLE OF THE NATIONAL CJD SURVEILLANCE UNIT (NCJDSU) DEPARTMENT OF NEUROPATHOLOGY, BEAUMONT HOSPITAL

4.1 **Where the suspected case is still alive**, the NCJDSU will:

- Arrange testing of CSF samples for 14-3-3 analysis if required/considered appropriate. (see page 7 for details)
- Notify the following of all possible cases of CJD to:
  - MOH in the area of residence of the case
  - IPTSE
- Notify the following of all probable cases of CJD to:
  - MOH in the area of residence of the case
  - IPTSE
  - Irish Blood Transfusion Service (IBTS)
- Notify the following of all probable/possible cases of vCJD to:
  - MOH in the area of residence of the case
  - IPTSE
  - IBTS

4.2 **Where the suspected case has died**, the NCJDSU will arrange for post mortem (PM) examination of the brain in order to obtain a definitive diagnosis.

4.3 **If a diagnosis of CJD or vCJD is confirmed** the NCJDSU will notify the following:

- Treating clinician
- Local coroner (where the patient is deceased)
- MOH in the area of residence of the case
- IPTSE
- IBTS
- Health Protection Surveillance Centre (HPSC) (anonymised)

4.4 **If CJD or vCJD is excluded as the diagnosis** the NCJDSU will advise the following:

- Treating clinician
- IBTS (to close case)
- MOH (to close case)
- Coroner (PM report) if the patient is deceased

SECTION 5: ROLE OF THE INCIDENT PANEL ON TSES (IPTSE)

5.1 To assist all those bodies responsible for the provision and delivery of healthcare to decide on the most appropriate action to take to handle incidents involving potential transmission of CJD and vCJD between patients through clinical interventions, including via reusable[10] invasive medical devices (RIMD[11]), tissues, organs and blood.

5.2 To:

- consider what information should be collected on patients who may have been exposed
- provide advice on what studies or follow-up may be needed
- advise Directors of Public Health and HITs on patient tracing and notification exercises where these are indicated; and
- provide advice on whether any other measures are needed to protect the wider public health.

10 **Definition**
Reusable invasive medical device: an invasive medical device that is designated or intended by the manufacturer as suitable for processing and reuse.

11 **Annex IX** of the **Medical Devices Directive (93/42/EEC) defines an invasive device as:**
A device which, in whole or in part, penetrates inside the body, either through a body orifice or through the surface of the body. A body orifice is defined as any natural opening in the body, as well as the external surface of the eyeball, or any permanent artificial opening, such as a stoma. The Directive also distinguishes a surgically invasive device as an invasive device which penetrates inside the body through the surface of the body, with the aid or in the context of a surgical operation. For the purposes of this Directive devices other than those referred to in the previous subparagraph and which produce penetration other than through an established body orifice, are treated as surgically invasive devices.

SECTION 6: ROLE OF THE HOSPITAL INVESTIGATION TEAM (HIT)

6.1 A Hospital Investigation Team (HIT) should be established in each hospital/healthcare facility attended by the patient where CJD or vCJD transmission may have occurred. If an incident is identified, the HIT should be chaired by the hospital's CEO/general manager and managed in line with HSE Safety Incident Management Policy.

6.2 Recommended members (guided by circumstances of each event):
Initially a small team is usually involved in the determination whether a CJD/vCJD incident has taken place.
If an incident is thought possible the following members are recommended:

- Hospital's CEO/general manager
- Treating consultant clinician
- Consultant surgeon/other clinician involved in any procedures undertaken
- MOH
- Hospital's infection prevention and control team including:
  - Consultant microbiologist
  - Decontamination coordinator
  - Infection prevention and control nurse
  - Central Decontamination Unit (CDU) / Endoscope Reprocessing Unit (ERU) personnel as indicated
- Risk manager
- Theatre manager
- Communications
- Others as required – IPTSE, General Practitioner (GP), NCJDSU, HPSC

6.3 Actions required of the HIT:

6.3.1 **Determine if a CJD or vCJD incident has occurred in their hospital**. Refer to "Algorithm for the identification of a CJD/vCJD Incident" for guidance (Appendix 8)

- **If no:** follow steps in either 6.3.2/6.3.3 as determined by diagnostic likelihood etc

- **If yes:**
  - Identify, locate and quarantine RIMD. It may be necessary to advise other hospitals or locations (e.g. manufacturing/maintenance company) of the need to quarantine RIMD (e.g. equipment was transferred, loaned, sent for repair or service)
  - Review the decontamination procedures and track and trace records which were followed for the RIMD concerned[12]
  - Complete Form B (CJD Incident Reporting form) (see Appendix 12) and forward this to the MOH / chair of the Case Investigation Team (CIT) (if established) and the IPTSE. **This form is used to report CJD incident** e.g. relevant surgical or other invasive procedures; blood donations.
  - Refer to Section 10. Managing the risk of possible exposure of patients following a surgical procedure
  - Contact the IPTSE who will advise on future use of RIMD and any follow-up of other exposed patients
  - Follow steps in either 6.3.2/6.3.3 as determined by diagnostic likelihood etc

12 HSE Decontamination RIMD Standards and recommended practices for Central Decontamination Units; Endoscope Reprocessing Units; Dental Central Decontamination Units & Local Decontamination Services are available at: https://www.hse.ie/eng/about/who/qid/quality-and-patient-safety-documents/decontamination/hse-standards-and-recommended-practices-for-endoscope-reprocessing-units.htm

Prevention of CJD and vCJD by Advisory Committee on Dangerous Pathogens' Transmissible Spongiform Encephalopathy (ACDP TSE) Subgroup: Annex E-Quarantining of surgical instruments https://www.gov.uk/government/uploads/system/uploads/attachment_data/file/209764/Annex_E_-_Quarantining_of_surgical_instruments.pdf
Annex F- Guidance on decontamination of flexible endoscopes for TSE infection prevention and control https://www.gov.uk/government/uploads/system/uploads/attachment_data/file/470292/ACDP_TSE_Annex_F_Oct_2015.pdf

6.3.2 If the patient has a confirmed/probable/possible diagnosis of CJD:

- Determine whether patient had received tissue or organ transplant
- Determine whether patient had donated tissue or organ
- If applicable, amend Form B accordingly and forward to the MOH/chair of the CIT (if established) and the IPTSE

6.3.3 If the patient has a confirmed/probable/possible diagnosis of vCJD:

- Determine whether this patient received any blood components e.g. whole blood, red blood cells, platelets, white cells (granulocytes or buffy coat preparations), plasma or cryoprecipitate in Ireland, the United Kingdom (UK) or France. For IBTS blood components, details of unit ID numbers of each blood component are required and dates of transfusion which should then be forwarded to IBTS
- Determine if this patient was treated with certain UK sourced plasma products
- Determine whether this patient had donated blood, tissues or organs
- Determine whether this patient had received tissue or organ transplant
- If applicable, amend Form B accordingly and forward to the MOH/chair of the CIT (if established) and the IPTSE

6.4 Case Investigation Team (CIT):

- If more than one healthcare facility is involved and multiple HITs have been established then a Case Investigation Team (CIT) may need to be set up by the MOH (see Section 7 and 8). This is most likely required in the case of incidents being identified. Each HIT should liaise closely with the Case Investigation Team (CIT)
- If only one healthcare facility has been identified then a CIT is probably not necessary and the full investigation can be carried out by the HIT in conjunction with the IPTSE

SECTION 7: ROLE OF THE LOCAL DEPARTMENT OF PUBLIC HEALTH – MEDICAL OFFICER OF HEALTH (MOH)

7.1 The MOH, in the area of diagnosis, will liaise with the MOH in the area of residence of the case. Practicalities of case management will guide which department takes the lead.[13]

7.2 The MOH in the lead Department of Public Health will:

- Review Form A (see Appendix 11) for completeness and seek clarifications if required
- Send any updated versions of Form A to the NCJDSU and the IPTSE
- Commence CIDR entry
- Determine which hospitals (including public, private, in this jurisdiction and in other jurisdictions) were attended by the case. This entails linking with clinicians involved including general practitioners(s) (current and past). Information may be required from family members. The treating clinician will usually be the liaison link with the family members
- If more than one hospital / healthcare facility is identified:
  - Establish a Case Investigation Team (CIT)- this is recommended particularly if an incident is identified
  - Contact the other hospitals involved and advise regarding the case and the need to establish and follow steps of a HIT
- Chair the Case Investigation Team (CIT) (see Section 8)
- If only one hospital / healthcare facility is identified:
  - Ensure MOH representation on the HIT

7.3 If the patient has a confirmed/probable/possible diagnosis of vCJD or if vCJD diagnosis remains unclear contact the IBTS (see Section 9)

7.4 If the patient has a confirmed or probable diagnosis of CJD contact the IBTS (see Section 9)

7.5 Update the IPTSE if a case's diagnosis changes e.g. if a patient's diagnosis changes from "diagnosis unclear" to "confirmed". This can be done on Form A.

13 Practicalities to consider include the location of any hospitals identified. Once there is a hospital incident, on site Consultant in Public Health Medicine involvement is recommended. More than one Department of Public Health may be involved in the overall investigation with one department taking the lead and chairing the CIT.

SECTION 8: ROLE OF THE CASE INVESTIGATION TEAM (CIT)

8.1 The key function of the CIT is to:

- Co-ordinate the overall investigation if more than one hospital / healthcare facility is involved

8.2 Members of CIT:

Initially a small team is usually involved in the determination whether a CJD/vCJD incident has taken place. If an incident is thought possible the following members are recommended:

- MOH (Chair)
- Representatives of the Hospital Investigation Teams (HITs)
- National Decontamination Lead
- Others as required: the case's GP(s), IPTSE, HPSC, NCJDSU, Senior Medical Officers, Surveillance Scientists, Communications

8.3 Specific actions of the CIT should include:

- Contact each hospital involved in patient care over the relevant period and advise the hospital CEO of diagnosis and the need to establish a HIT

If a CJD/VCJD incident is considered likely:

- Review and collate all data received in Form B (see Appendix 12) from each HIT and forward the combined information to the IPTSE
- Assess (with the IPTSE) the possible follow-up of contacts following a CJD or vCJD incident. Irreversible actions such as permanent disposal of RIMD and notification of subsequently exposed patients ought not to be conducted until the CJD diagnosis or risk status has been confirmed. Irreversible actions such as permanent disposal of RIMD and notification of subsequently exposed patients ought not to be conducted until the CJD diagnosis or risk status has been confirmed AND with the agreement of the IPTSE.
- Co-ordinate (with the IPTSE) the overall communications strategy if more than one hospital / healthcare facility is involved
- Develop and implement a process to contact patients if this is deemed necessary (see Section 10). This should be done in conjunction with the IPTSE and the HITs.

SECTION 9: ROLE OF THE IRISH BLOOD TRANSFUSION SERVICE (IBTS)

9.1 If the patient has a confirmed/probable/possible diagnosis of **v**CJD the role of the IBTS is to:

- Determine whether this patient had donated blood or blood components

If yes, then to advise the MOH and the IPTSE. Work with the IPTSE and HIT/CIT on management/lookback/ communications etc

- Recall and withdraw from use any in-date blood components
- If a donor, permanently exclude from donating blood or blood components

9.2 If the patient has a confirmed or probable diagnosis of **Sporadic or Familial** CJD/other TSE the role of the IBTS is to:

- Determine whether this patient had donated blood or blood components. Recall and withdraw from use any in-date blood components
- If a donor, permanently exclude from donating blood or blood components

9.3 If the patient has a confirmed or probable diagnosis of **Iatrogenic** CJD/other TSE the role of the IBTS will be determined on an individual case basis.

SECTION 10: MANAGING THE RISK OF POSSIBLE EXPOSURE OF PATIENTS FOLLOWING A SURGICAL PROCEDURE

10.1 Refer to the Algorithm for the Identification of a CJD or vCJD incident (Appendix 8).[14]

10.2 Managing the risk of possible exposure of other patients is a shared role of the HIT, CIT (if established) and IPTSE

10.3 The number of contacts (exposed patients) depends on:

- The type of CJD the index patient has
- Why the index patient is at increased risk of CJD or vCJD
- The infectivity of the tissues involved in the procedure
- The number of times the implicated RIMD have been used since the index patient's procedure

Appendix 7 outlines the Public Health Actions required following the report of a new case of CJD or a person at increased risk of CJD.[15]

10.4 It is clear that each exposure will be unique and will be managed with an incident dependent approach. Issues for consideration include:

- Identification of the patients on whom the RIMD were used subsequent to the index patient's procedure. This information is to be shared and discussed with the MOH/Chair of the CIT and the IPTSE
- The IPTSE will advise on what contacts, if any, should be considered at risk for public health purposes and informed of this risk bearing in mind the following issues:
  - Confidentiality of the process
  - Communication strategy for patients and staff
  - Contacting patients deemed to be exposed
  - Advice and counselling options for patients
  - Patient data that should be collected
  - How collected data should be stored and by whom
  - Arrangement of any necessary follow up- most importantly how these individuals will be "flagged" in the health system to enable identification if they present subsequently for procedures/surgery
  - Each HIT should retain all incident information, including names of patients possibly exposed but not considered to be 'at-risk'.

14 Please refer to https://www.gov.uk/government/organisations/public-health-england to check for updated guidance documents

15 Inserted are Tables 1-5 of the Public Health England and Health Protection Scotland document *Public Health Action Following a Report of a New Case of CJD or a Person at Increased Risk of CJD* (pgs 11 & 12) https://assets.publishing.service.gov.uk/government/uploads/system/uploads/attachment_data/file/474338/CJD_public_health_action_new_case_301015.pdf

SECTION 11: CASES CLASSIFIED AS "DIAGNOSIS UNCLEAR"

11.1 Some cases are classified as "diagnosis unclear" i.e. the diagnostic criteria for confirmed/probable/possible CJD or vCJD are not met, nor is there a reasonable alternative diagnosis. CJD therefore remains as a differential diagnosis.

11.2 All efforts should be made by the treating clinician to promptly ascertain the likelihood of a CJD diagnosis and ideally before any procedures are carried out.

- An experienced neurologist should be directly involved in the case assessment.
- Review of all data is required including clinical information, CSF, EEG and MRI.
- Where uncertainty remains the treating clinician should liaise as soon as possible with the IPTSE neurologist. The case may require discussion with the IPTSE.

11.3 The relevant steps of this protocol should commence by the treating clinician unless advised otherwise by the IPTSE i.e. recommence at section 3- completing Form A etc.

11.4 As part of the protocol, it should be ascertained **whether the patient had any invasive procedures involving high or medium level risk tissues over a past timeline**. This timeframe will be dependent on the individual circumstances and may require discussion with the IPTSE.

See Algorithm for the identification of a CJD/TSE Surgical Incident in Appendix 8 .

If yes, follow relevant steps including quarantine of RIMD.

11.5 Where vCJD is being considered contact the Irish Blood Transfusion Service (IBTS) and advise re diagnostic query.

11.6 When the diagnosis changes the relevant clinician should advise the MOH of this change in category and of the definitive diagnosis and an updated Form A should be submitted to the MOH. The IPTSE and the IBTS should then be informed by the MOH of these updates

- Cases classified as "diagnosis unclear" may move into the following categories:
  - **"Confirmed/probable/possible diagnosis of CJD"**: In this instance the relevant steps in this protocol should be followed.
  - **"CJD thought unlikely"** –i.e. 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. **There is no need for CJD Public Health precautions in relation to this case.**
  - **"Definitely not CJD"** - An alternative diagnosis, that rules-out CJD or other TSE, has been made and confirmed. **There is no need for CJD Public Health precautions in relation to this case.**

SECTION 12: (AFTER DEATH) PROTOCOL FOR THE REFERRAL OF AUTOPSIES TO THE NCJDSU, BEAUMONT HOSPITAL (PLEASE SEE APPENDIX 9)

12.1 The diagnosis of probable or possible CJD can be made during the clinical illness in individual patients but usually the only way to establish the definitive diagnosis is by performing an autopsy examination of the brain. All efforts should be made for post mortem examination of brain tissue by the NCJDSU (Beaumont Hospital).

12.2 All cases of individuals who have died with, or as a result of, suspected CJD/vCJD in Ireland must be reported to the local coroner by the treating clinician. It is at the coroner's discretion whether or not a post mortem takes place. In the event that the coroner does not direct that a post mortem examination occur, the clinician responsible for the patient's care at the time of death may request consent for autopsy from the next of kin. The clinician should not request consent for an autopsy in advance of notifying the coroner. In both instances the treating clinicians should inform the family that the remains must be transferred to Beaumont Hospital for autopsy.

12.3 After reporting the case to the coroner, the neuropathologists in Beaumont Hospital should be contacted by the treating clinician. The neuropathology secretary can be contacted at 01-8092631, or if it is out of hours, this is done by contacting the Beaumont Hospital switch 01-8093000 and asking for the neuropathologist on call.

12.4 **All appropriate documentation must be completed and sent to the NCJDSU (Beaumont Hospital) prior to or immediately after patient referral.** These documents are comprised of Form A (appendix 11), Form C example of a CJD post mortem information form (appendix 13) and a form relating to cremation of body parts.[16]

12.5 The referring hospital / nursing home staff must obtain Garda identification, which is then transferred to the undertaker responsible for transporting the body to Beaumont Hospital (i.e. the undertaker looking after the family).

12.6 Transportation of the remains with appropriate identification (i.e. wrist or ankle badges or both) and hospital notes to Beaumont Hospital is the responsibility of the referring hospital / nursing home. The body should be transported in a body bag to protect against accidental seepage of body fluids following death.

12.7 At the NCJDSU, western blot for prion protein is carried out on several brain regions by the Medical Scientist. If this is positive, the brain is pre-treated with formic acid for further analysis and is then retained in formalin until the autopsy report has been completed. If the western blot is negative then routine analysis is carried out.

12.8 Following neuropathological examination the brain can be returned to the undertaker dealing with the family for either burial or cremation. Alternatively it can be disposed of respectfully by Beaumont Hospital. The family need to decide which is their preference.

12.9 The treating clinician must inform the family that the brain will be retained during post mortem examination and can be returned to the family undertaker or disposed of respectfully by Beaumont Hospital following diagnosis. Please see Appendix 13 Form C and form relating to cremation of body parts.[16]

12.10 The final report of the autopsy examination is issued to the coroner. If the post mortem is not carried out under a coroner's jurisdiction the post mortem report will be sent to the treating clinician and to the MOH in the region of the patient's death as well as the MOH in Dublin. The following will also be notified of the case: IBTS, HPSC, IPTSE.

16 Example of form relating to the cremation of body parts https://www.glasnevintrust.ie/funeral-services/downloads/

SECTION 13: FUNERAL ARRANGEMENTS FOR CJD/VCJD PATIENTS/PATIENTS "AT INCREASED RISK" OF ANY FORM OF CJD

13.1 Funeral arrangements for a person who dies with CJD or vCJD are the same as for other individuals; with the exception that embalming is not recommended.

13.2 When the diagnosis of CJD/vCJD is known or suspected, it is advisable to avoid embalming procedures. When embalming is required, then it should only be carried out in specialist facilities fit for the purpose of handling TSE cases.

13.3 Viewing the deceased: Relatives, friends or carers of the deceased may wish to view or have some final contact with the deceased. Such viewing and possible superficial contact, such as touching or kissing, need not be discouraged even if a post-mortem has taken place. Body bags may be rolled down temporarily to allow superficial contact; there is no need to deny the relatives, friends or carers this opportunity if a post-mortem examination has been performed.

Answers to some commonly asked questions[17]

13.4 Are there any risks to relatives in viewing the body of a patient who has died with CJD?
There is no evidence that CJD can be passed from one person to another by contact with the skin or hair. Therefore, the body bag can be opened to allow relatives to view the body, and, if they wish, have contact with the deceased. Such viewing and possible superficial contact, such as touching or kissing, need not be discouraged even if a post-mortem has taken place.

13.5 If an autopsy has been performed are there any additional risks to viewing the body of a patient who has died of CJD?
No, as above, the body bag can be opened to allow relatives to view the body, and, if they wish, have contact with the deceased, with no additional risk to either staff or relatives.

13.6 Are there any risks to relatives in dressing the body and washing the hair of a patient who has died of CJD?
As above, there is no evidence that CJD can be passed from one person to another by contact with the skin or hair. Therefore, the body bag can be opened to allow relatives to dress the body and wash the hair.

13.7 If an autopsy has been performed are there any additional risks to dressing the body and washing the hair of a patient who has died of CJD?
If an autopsy has been performed, dressing of the body and washing of the hair may be performed by relatives under the supervision of mortuary staff or a funeral director, using standard infection control measures to minimise risk.

13.8 Are there any risks involved in transporting the body of a patient who has died with CJD?
Precautions are required for the transport of people who have died with CJD. The body should be transported in a body bag to protect against accidental seepage of body fluids following death.

13.9 Are special burial or cremation arrangements required for a patient who has died with CJD?
No special arrangements are needed for burial or cremation of a patient with known or suspected CJD. Regarding embalming please refer to 13.1.

17 Guidance for undertakers when dealing with patients who have died with CJD
http://www.cjd.ed.ac.uk/sites/default/files/Guidance%20for%20Undertakers.pdf

APPENDIX 1:

Draft Pre-operative CJD/vCJD Risk assessment FOR HIGH RISK TISSUES form

Name_____

MRN_____

DOB_____

WARD_____

COMPLETE OR ATTACH AN ADDRESSOGRAPH LABEL

To be completed prior to surgery/ endoscopy/invasive procedures where contact with high risk TSE tissue* is likely

1. Does the patient have a confirmed TSE?	YES (Neuropathological / immunocytochemical confirmation)	NO
2. Does the patient have a clinically suspected TSE?	YES (patient fulfils diagnostic criteria for definite, probable or possible TSE/CJD/vCJD[1] **or** patient has neurological disease of unknown aetiology and CJD is being actively considered)	NO
3. Has the patient been advised that he/she is at increased risk of CJD/vCJD? PTO for list.	YES	NO
4. Does the patient have a history of CJD/other prion disease in his/her family? E.g. two or more relatives with a confirmed TSE; the patient or any of their relatives have a known genetic risk of TSE identified on specific genetic testing?	YES	NO
5. Has the patient had growth hormone or gonadotrophin treatment (used prior to 1987)?	YES	NO
6. Has the patient had brain or spinal cord surgery prior to July 1996? (Individuals who underwent intradural brain or intradural spinal surgery before July 1996 might have received a graft of human-derived dura mater)	YES	NO
7. Has the patient received blood from 300 or more UK donors since Jan 1990?	YES	NO
8. Has the patient received UK sourced plasma products between 1990 and 2001?	YES	NO

If the patient answered YES to any question please notify Infection Prevention and Control immediately AND prior to proceeding with the procedure. Refer to Infection Control policy on TSE/CJD.

Signature (person completing form):

_____

Position:

_____

Print Name:

_____

Date:

_____

***high risk TSE tissue includes:**
Brain
Spinal cord
Dura mater
Cranial nerves, specifically:
- the entire optic nerve
- the intracranial components of the other cranial nerves

Cranial nerve ganglia
Posterior eye, specifically:
- posterior hyaloid face
- retina
- retinal pigment epithelium
- choroid
- subretinal fluid
- optic nerve

Pituitary gland

Patients identified to be at increased risk include:

Related to blood transfusions

- People who have received blood or blood components from someone who went on to develop vCJD
- People who have given blood or blood components to someone who went on to develop vCJD
- People who have received blood or blood components from someone who has also given blood or blood components to a patient who went on to develop vCJD
- People who have received blood or blood components from 300 or more UK donors since 1990 (additional information on how this group is defined can be found in the ACDP TSE guidance FAQ document.[18]

Related to surgery

- People who have had surgery using instruments that had been used on high or medium level risk tissues of someone who developed CJD
- People who underwent an intradural neurosurgical or intradural spinal procedure **before July 1996 who received (or might have received) a graft of human-derived dura mater.**
- People who have received an organ or tissue from a donor infected with CJD or at increased risk of CJD

Related to other medical care

- **Treatment with certain UK sourced plasma products between 1990 and 2001 (inclusive)**
- People who have been treated with growth hormone **from UK sourced human pituitary glands (before 1987)**
- People who have been treated with gonadotrophin sourced **from human pituitary glands for fertility treatment (before 1987)**
- People who have been told by a specialist that they have a risk of developing the genetic form of CJD

18 Advisory Committee on Dangerous Pathogens Transmissible spongiform encephalopathy agents: safe working and the prevention of infection Frequently asked questions. www.gov.uk/government/uploads/system/uploads/attachment_data/file/271414/Frequently_asked_questions.pdf

APPENDIX 2:

Assessment to be carried out before undergoing surgery or neuro-endoscopy which may involve contact with tissues of potentially high level TSE infectivity ("high risk tissues") to identify patients with, or at increased risk of, CJD or vCJD

(Adaption of Transmissible Spongiform Encephalopathy Agents: Safe Working and the Prevention of Infection: Annex J[19])

Summary of advice

It is recommended that patients undergoing surgery or neuro-endoscopy which may involve contact with tissues of potentially high level TSE infectivity ("high risk tissues") should, through a set of detailed questions, be assessed for their possible CJD/vCJD risk exposure. These questions are outlined in Tables J1 and J2 and paragraph J1.

Tissues assumed or proven to have **high** level infectivity for CJD or vCJD are:

Brain

Spinal cord

Cranial nerves, specifically:
- the entire optic nerve
- only the intracranial components of the other cranial nerves

Cranial nerve ganglia

Posterior eye, specifically:
- posterior hyaloid face
- retina
- retinal pigment epithelium
- choroid
- subretinal fluid
- optic nerve

Pituitary gland

Assessments prior to surgery and neuro-endoscopy which may involve contact with high risk tissue

J1. N.B. These recommendations are applicable to those assessing patients in neurosurgical and ophthalmic surgical departments for intradural and posterior ophthalmic surgical procedures. With regards to endoscopy, these recommendations are applicable to those assessing patients for intradural neuro-endoscopic procedures.

Procedures should not be delayed whilst information is being collected, and clinicians should be careful not to prejudice overall patient care. **Please consider single use instruments in these circumstances.**

All patients about to undergo any elective or emergency surgical or endoscopic procedures that involve contact with high risk tissues should be asked these questions:

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

19 https://webarchive.nationalarchives.gov.uk/20120503130558/http://www.dh.gov.uk/prod_consum_dh/groups/dh_digitalassets/@dh/@ab/documents/digitalasset/dh_123586.pdf

Table J1

Question 1: "Have you ever been notified that you are at increased risk of CJD or vCJD?"	
Patient's response	**Actions to take following the patient's response to the above question**
No	Continue with following questions
Yes	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 tissues and the local infection prevention and control team should be consulted for advice. Part 4 of the Public Health England Guidance (https://www.gov.uk/government/uploads/system/uploads/attachment_data/file/427854/Infection_controlv3.0.pdf) provides advice on the precautions to be taken during the treatment of patients with or at increased risk of CJD or vCJD, and Annex F (https://www.gov.uk/government/uploads/system/uploads/attachment_data/file/470292/ACDP_TSE_Annex_F_Oct_2015.pdf) provides information on endoscopic procedures. The patient's response should be recorded in their medical notes for future reference.
Unable to respond	Please refer to the additional recommendations for high risk procedures from paragraph J2 onwards, with particular reference to paragraphs J2 – J4.

Table J2

Table J2	Further questions to patient	Notes to clinician
Q2	Have you a history of CJD or other prion disease in your family? If yes, please specify.	Patients should be considered to be at 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 disease. If the patient answers No or Not Certain to all 3 of these questions then surgery can proceed with normal infection prevention and control procedures.
Q3	Have you ever received growth hormone or gonadotrophin treatment? If yes, please specify: i) whether the hormone was derived from human pituitary glands ii) the year of treatment iii) which country the treatment was received in?	Recipients of hormone derived from human pituitary glands, e.g. growth hormone or gonadotrophin, have been identified as at increased risk of sporadic CJD. In Ireland, the use of human-derived growth hormone was discontinued by 1987 but human-derived products may have continued to be used in other countries. In Ireland the use of human-derived gonadotrophin was discontinued by 1987 but may have continued in other countries after this time.
Q4	Have you had surgery on your brain or spinal cord prior to July 1996?	Individuals who underwent intradural brain or intradural spinal surgery before July 1996 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).
Q5	Have you received blood from 300 or more UK donors since Jan 1990?	
Q6	Have you received UK sourced plasma products between 1990- 2001?	

Table J3. The actions to be taken following the patient's response to the above questions are:

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 1-6	Further investigation 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 brought to the attention of the IPTSE who will then form an opinion. Only then should a patient be advised of their risk status. This assessment should be recorded in the patient's medical notes for future reference. If the patient is found to be at increased risk of CJD or vCJD following investigation, or the 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 the local infection prevention and control team should be consulted for advice. Part 4 provides advice for the precautions to be taken during the treatment of patients with or at increased risk of CJD or vCJD, and Annex F provides information on neuro-endoscopic procedures.
Unable to respond	See paragraphs J2 – J4 below for advice.

Emergency surgery or neuro-endoscopy which may involve contact with high risk tissue

J2. 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 prior to the surgery or neuro-endoscopy.

J3. If the family member, or someone close to the patient, is not able to provide a definitive answer to the CJD risk questions, the surgery or neuro-endoscopy should proceed but all instruments should be quarantined following the procedure - refer to Annex E and F.

The patient's GP should be contacted after the surgery or neuro-endoscopy, and enquiries made as to whether the patient is at increased risk of CJD/vCJD according to the CJD risk questions.

Table J4. The actions to be taken following the GP's response to the CJD risk questions

GP's response	Action
No to all questions	The instruments can be returned to routine use after undergoing normal decontamination processes.
Yes to any of questions 1-6	Further investigation into the nature of the patient's CJD risk should be undertaken, and the patient's CJD risk confirmed or rejected. Confirmation or rejection of CJD risk should be recorded in the patient's medical notes for future reference. If the patient is found to be at increased risk of CJD or vCJD following investigation then the quarantine instruments should be destroyed or retained for exclusive use of the patient. Refer to Appendix 5.
Uncertain about any of questions 1-6	The instruments should be kept in quarantine. The local infection prevention and control team should carry out a risk assessment, and they should involve the local Consultant in Public Health Medicine and IPTSE in this process. The outcome of the risk assessment should determine whether or not to return the instruments to routine use.

Additional actions to be taken during pre-surgery assessment for CJD risk

J4. In addition to asking the patient CJD/vCJD risk questions, the following actions should also be carried out before any surgical or endoscopic procedure involving contact with high risk tissue. The clinician undertaking the pre-surgery assessment should:

- Check the patient's medical notes and/or referral letter for any mention of CJD or vCJD status
- Consider whether there is a risk that the patient may be showing the early signs of CJD or vCJD, i.e. consider whether the patient may have an undiagnosed neurological disease involving cognitive impairment or ataxia

APPENDIX 3:

Draft information for patients undergoing surgery or neuro-endoscopy on high risk tissues

Part of your routine assessment before surgery includes some questions to find out whether you could have an increased risk of Creutzfeldt-Jakob disease (CJD). We will ask you:

Have you ever been notified that you are at risk of CJD or vCJD for public health purposes?

Have you any history of CJD or other prion disease in your family?

Have you ever received growth hormone or gonadotrophin treatment?

Have you had surgery on your brain or spinal cord at any time in the past?

Have you received blood transfusions or plasma products in the past?

What is CJD?
Creutzfeldt-Jakob disease (CJD) is a very rare brain disorder. CJD is thought to be caused by the build up in the brain of an abnormal form of a protein called a 'prion'. Unfortunately CJD is fatal, and as yet there is no known cure. There are different types of CJD, including variant CJD (vCJD). vCJD is caused by eating meat from cows infected with BSE.

How can CJD spread from person to person?
A person who is infected with CJD may have abnormal prion protein in their body for years before becoming ill. If that person has an operation, or donates blood, tissues or organs, during that time, the abnormal prion protein that causes CJD could spread to other patients.

Why are we asking you about CJD before your operation?
The abnormal prion protein that causes CJD is very hard to remove or destroy. If surgical instruments are used on a patient who is infected with CJD they may still have prion protein on them, even after they have been properly washed and disinfected. They could then spread CJD to other patients. This is particularly important for operations on the brain, spinal cord and the back of the eye as these parts of the body contain the largest amount of abnormal prion protein.

What have these questions got to do with CJD?
CJD has been spread in several ways and different groups of people may have an increased risk of CJD.

We ask whether there is anyone in your family who has had CJD because some types of CJD can be inherited. These types of CJD are caused by faulty genes and may be passed from parent to child.

We ask whether you have had surgery on the brain or spinal cord because some of these operations used to use grafts of 'dura mater' (the tough lining round the brain and spinal cord). Some of these grafts have been linked to CJD infection - these grafts are no longer used.

We ask whether you have been treated with growth hormone or gonadotrophin infertility treatment because these used to be prepared from pituitary glands. Some of these hormone treatments have been linked to CJD infection - these hormones are no longer used.

We ask whether you have had a large number of blood transfusions as this could be related to an increased risk of variant CJD (vCJD). vCJD is the type of CJD which is caused by eating meat from cows infected with BSE. vCJD can be spread through blood transfusions.

What happens if I answer 'Yes' to any of these questions?
If you answer 'Yes' to any of these questions, medical staff will now examine your medical records in more detail to determine whether or not you may have an increased risk of CJD.

What will happen then?
If you do have an increased risk of CJD special precautions will be taken with the surgical instruments used in your operation. Your GP will be informed.

Please remember that the overall risk of CJD spreading by these routes is generally very low. These questions are an extra measure to prevent CJD spreading through surgery. This should not affect the medical care you receive now or in the future.

Can I have a blood test to see if I am infected with CJD?
Unfortunately there is no blood test available yet which could show if you have CJD.

Where can I find out more?
The following organisations offer further information. Most of these organisations are not based in the Republic of Ireland.

Health Protection Surveillance Centre: www.hpsc.ie

CJD Support Network website: www.cjdsupport.net

NHS Choices: www.nhs.uk/conditions/Creutzfeldt-Jakob-disease/Pages/Introduction.aspx

National Prion Clinic website: www.nationalprionclinic.org/

APPENDIX 4:
DIAGNOSTIC CRITERIA[20]

1. SPORADIC CJD

1.1 DEFINITE
Progressive neurological syndrome **AND** Neuropathologically **or** immunocytochemically **or** biochemically confirmed

1.2 PROBABLE
1.2.1 I + 2 of II and III
OR
1.2.2 I + 2 of II and IV
OR
1.2.3 I + 2 of II and positive 14-3-3
OR
1.2.4 Progressive neurological syndrome and positive RT-QuIC in CSF or other tissues

1.3 POSSIBLE
I + 2 of II + duration < 2 years

I Rapidly progressive cognitive impairment

II A Myoclonus
B Visual or cerebellar problems
C Pyramidal or extrapyramidal features
D Akinetic mutism

III Typical EEG (Generalised periodic complexes)

IV High signal in caudate/putamen on MRI brain scan or at least two cortical regions (temporal, parietal, occipital) either on DWI or FLAIR"

2. ACCIDENTALLY TRANSMITTED TSE

2.1 DEFINITE
Definite CJD with a recognised iatrogenic risk factor (see box)

2.2 PROBABLE
2.2.1 Progressive predominant cerebellar syndrome in human pituitary hormone recipients
2.2.2 Probable CJD with recognised iatrogenic risk factor (see box)

2.3 POSSIBLE
Possible CJD with a recognised risk factor
(agreed and EURO meeting Bled, 2006)

RELEVANT EXPOSURE RISKS FOR THE CLASSIFICATION AS IATROGENIC CJD

The relevance of any exposure to disease causation must take into account the timing of the exposure in relation to disease onset

- Treatment with human pituitary growth hormone, human pituitary gonadotrophin or human dura mater graft.
- Corneal graft in which the corneal donor has been classified as definite or probable human prion disease.
- Exposure to neurosurgical instruments previously used in a case of definite or probable human prion disease.

This list is provisional as previously unrecognised mechanisms of human prion disease may occur

20 DIAGNOSTIC CRITERIA OF THE (UK) NATIONAL CJD RESEARCH & SURVEILLANCE UNIT (NCJDRSU)
http://www.cjd.ed.ac.uk/sites/default/files/criteria_0.pdf

DIAGNOSTIC CRITERIA CONTINUED[7]

3. GENETIC TSE

3.1 DEFINITE

3.1.1 Definite TSE + definite or probable TSE in 1st degree relative

3.1.2 Definite TSE with a pathogenic PRNP mutation (see box)

3.2 PROBABLE

3.2.1 Progressive neuropsychiatric disorder + definite or probable TSE in 1st degree relative

3.2.2 Progressive neuropsychiatric disorder + pathogenic PRNP mutation (see box)

- **PRNP MUTATIONS ASSOCIATED WITH GSS NEUROPATHOLOGICAL PHENOTYPE**
  P102L, P105L, A117V, G131V, F198S, D202N, Q212P, Q217R, M232T, 192 bpi
- **PRNP MUTATIONS ASSOCIATED WITH CJD NEUROPATHOLOGICAL PHENOTYPE**
  D178N-129V, V180I, V180I+M232R, T183A, T188A, E196K, E200K, V203I, R208H, V210I, E211Q, M232R, 96 bpi, 120 bpi, 144 bpi, 168 bpi, 48 bp*del*
- **PRNP MUTATIONS ASSOCIATED WITH FFI NEUROPATHOLOGICAL PHENOTYPE**
  D178N-129M
- **PRNP MUTATION ASSOCIATED WITH VASCULAR PRP AMYLOID**
  Y145s
- **PRNP MUTATIONS ASSOCIATED WITH PROVEN BUT UNCLASSIFIED PRION DISEASE**
  H187R, 216 bpi
- **MUTATIONS ASSOCIATED WITH NEURO-PSYCHIATRIC DISORDER BUT NOT PROVEN PRION DISEASE**
  I138M, G142S, Q160S, T188K, M232R, 24 bpi, 48 bpi, 48 bpi + nucleotide substitution in other octapeptides

(additional list of mutations below)

4. vCJD

4.1 DEFINITE

1A **and** neuropathological confirmation of vCJD[e]

4.2 PROBABLE

4.2.1 I **and** 4/5 of II **and** IIIA **and** IIIB

4.2.2 I **and** IV A[d]

4.3 POSSIBLE

I **and** 4/5 of II **and** III A

I A Progressive neuropsychiatric disorder
B Duration of illness > 6 months
C Routine invesitgations do not suggest an alternative diagnosis
D No history of potential iatrogenic exposure
E No evidence of a familial form of TSE

II A Early psychiatric symptoms[a]
B Persistent painful sensory symptoms[b]
C Ataxia
D Myoclonus or chorea or dystonia
E Dementia

III A EEG does not show the typical appearance of sporadic CJD[c] in the early stages of illness
B Bilateral pulvinar high signal on MRI scan

IV A Positive tonsil biopsy[d]

a depression, anxiety, apathy, withdrawal, delusions.

b this includes both frank pain and/or dysaesthesia.

c the typical appearance of the EEG in sporadic CJD consists of generalised triphasic periodic complexes at approximately one per second. These may occasionally be seen in the late stages of variant CJD.

d tonsil biopsy is **not** recommended routinely, nor in cases with EEG appearances typical of sporadic CJD, but may be useful in suspect cases in which the clinical features are compatible with vCJD and MRI does not show bilateral pulvinar high signal.

e spongiform change and extensive PrP deposition with florid plaques throughout the cerebrum and cerebellum.

ADDITIONAL LIST OF MUTATIONS

- **PRNP MUTATIONS WITHOUT CLINICAL AND NEUROPATHOLOGICAL DATA**
  T188R, P238S
- **PRNP POLYMORPHISMS WITH ESTABLISHED INFLUENCE ON PHENOTYPE**
  M129V
- **PRNP POLYMORPHISMS WITH SUGGESTED INFLUENCE ON PHENOTYPE**
  N171S, E219K, 24 bpdeletion
- **PRNP POLYMORPHISMS WITHOUT ESTABLISHED INFLUENCE ON PHENOTYPE**
  P68P, A117A, G124G, V161V, N173N, H177H, T188T, D202D, Q212Q, R228R, S230S

APPENDIX 5:

MANAGING POTENTIALLY CONTAMINATED REUSABLE INSTRUMENTS AND EXPOSED PATIENTS

NB: Assess (with the IPTSE) before any irreversible actions such as permanent disposal of RIMD and notification of subsequently exposed patients

Tables 1-5[20] set out the summary of actions to be taken to manage potentially contaminated instruments and patients exposed to a risk of CJD. Actions required are dependent on:

i) The CJD type the index patient has or why the index patient is at an increased risk of CJD

ii) The CJD infectivity of the tissues involved in the index patient procedure

iii) The number of times the implicated instruments have been used since the index patient procedure*

Use the algorithm below to identify the table of public health actions relevant to the index patient under investigation.

What type of CJD does the patient have or what is their increased risk status?

Symptomatic	Definite, probable or possible sporadic, inherited or iatrogenic CJD	Table **1**
	Variant CJD	Table **2**
	Unclear diagnosis	Table **5**
	At increased risk of inherited prion disease	Table **1**
	At increased risk of variant CJD through receiving blood from a donor who later developed variant CJD	Table **2**
Asymptomatic	At increased risk of variant CJD through other blood exposures or through treatment with UK sourced plasma products between 1990 and 2001	Table **3**
	At increased risk of iatrogenic CJD (other than variant CJD)	Table **4**

*** "Use" for the purposes of this document is defined as use of the instrument(s) on a patient followed by decontamination to an approved standard.**

20 Tables 1-5 of the recent Public Health England and Health Protection Scotland document *Public Health Action Following a Report of a New Case of CJD or a Person at Increased Risk of CJD* (pgs 11 & 12) https://www.gov.uk/government/publications/cjd-public-health-action-following-report-of-new-case-or-person-at-increased-risk.

TABLE 1: PUBLIC HEALTH ACTIONS REQUIRED FOR:

- **Symptomatic patients with Sporadic CJD / inherited prion disease / iatrogenic CJD* - definite/probable/ possible**
- **Asymptomatic patients at increased risk of inherited prion disease**

The following applies where an invasive procedure has been carried out.

Tissue involved in procedure	Action for instruments is determined by the number of cycles of use and decontamination they have already been through since used on the index patient				Patients exposed to instruments/RIMD
	Action for surgical instruments/**RIMD** by number of uses to date		Action for flexible endoscopes** by number of uses to date		
High infectivity • Brain • Spinal cord • Cranial nerves • Cranial ganglia • Posterior eye • Pituitary glands	Fewer than 20 uses Destroy or retain for exclusive use on this patient	More than 20 uses Reprocess & return to use	Fewer than 20 uses Destroy or retain for exclusive use on this patient	More than 20 uses Destroy or retain for exclusive use on this patient	10 patients subsequently exposed to instruments/RIMD in contact with high infectivity tissues should be traced and notified
Medium infectivity • Spinal ganglia • Olfactory epithelium**	Fewer than 10 uses Destroy or retain for exclusive use on this patient	More than 10 uses Reprocess & return to use	Fewer than 10 uses Destroy or retain for exclusive use on this patient	More than 10 uses Destroy or retain for exclusive use on this patient	2 patients subsequently exposed to instruments/RIMD in contact with medium infectivity tissues should be traced and notified
Low infectivity All other tissues not listed above	Reprocess & return to use	Reprocess & return to use	Reprocess & return to use	Reprocess & return to use	No patients should traced and notified

*iatrogenic CJD as a result of receiving human derived growth hormone, gonadotropin or a dura mater graft (excludes patients with iatrogenically acquired variant CJD)

** The olfactory epithelium is normally located deep within the nasal turbinates and normal nasal endoscope procedures do not reach the olfactory epithelium. However its distribution varies between individuals. 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 such contamination cannot be excluded, take precautions appropriate for medium infectivity tissues.

Note: before an instrument is quarantined it should be first decontaminated to the required standard (https://www.hse.ie/eng/about/who/qid/nationalsafetyprogrammes/decontamination/decontamination-of-rimd.html) for additional information refer to Annex E Quarantining of surgical instruments guidance and Annex C - General principles of decontamination and waste disposal https://assets.publishing.service.gov.uk/government/uploads/system/uploads/attachment_data/file/427855/Annex_C_v3.0.pdf

NB: Single use biopsy forceps should be used in all patients

TABLE 2: PUBLIC HEALTH ACTIONS REQUIRED FOR:

- **Symptomatic patients with vCJD-definite/probable/possible**
- **Asymptomatic patients at increased risk of vCJD through receiving blood from a donor who later developed vCJD***

A greater range of medium risk tissues should be considered during the risk assessment.

The following applies where an invasive procedure was carried out.

Tissue involved in procedure	Action for instruments/RIMD is determined by the number of cycles of use and decontamination they have already been through since used on the index patient				Patients exposed to instruments/RIMD
	Action for surgical instruments/RIMD by number of uses to date		Action for flexible endoscopes** by number of uses to date		
High infectivity • Brain • Spinal cord • Cranial nerves • Cranial ganglia • Posterior eye • Pituitary glands	Fewer than 20 uses Destroy or retain for exclusive use on this patient	More than 20 uses Reprocess & return to use	Fewer than 20 uses Destroy or retain for exclusive use on this patient	More than 20 uses Destroy or retain for exclusive use on this patient	10 patients subsequently exposed to instruments/RIMD in contact with high infectivity tissues should be traced and notified
Medium infectivity • Spinal ganglia • Olfactory epithelium** • Tonsil, adenoids • Appendix • Spleen • Thymus • Adrenal gland • Lymph nodes & gut-associated lymphoid tissues (including the rectum)	Fewer than 10 uses Destroy or retain for exclusive use on this patient	More than 10 uses Reprocess & return to use	Fewer than 10 uses Destroy or retain for exclusive use on this patient	More than 10 uses Destroy or retain for exclusive use on this patient***	2 patients subsequently exposed to instruments/RIMD in contact with medium infectivity tissues should be traced and notified
Low infectivity All other tissues not listed above	Reprocess & return to use	Reprocess & return to use	Reprocess & return to use	Reprocess & return to use	No patients should traced and notified

*Or theoretically also through receiving tissue or organs donated by a patient who later developed variant CJD (no known occurrences to date)

** The olfactory epithelium is normally located deep within the nasal turbinates and normal nasal endoscope procedures do not reach the olfactory epithelium. However its distribution varies between individuals. 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 such contamination cannot be excluded, take precautions appropriate for medium infectivity tissues.

***Flexible gastrointestinal endoscopes may be suitable for refurbishment by their manufacturers/distributors to allow their return to later use. This refurbishment process may be considered as an alternative to quarantining the instrument if a flexible gastrointestinal endoscope has beenused in the performance of an invasive procedure in patients at risk of vCJD because they received blood from a donor who later developed vCJD. Refurbishment is not available for endoscopes that have been used for invasive endoscopy in patients with definite or probable vCJD. The decision to undertake refurbishment will be made on a case by case basis by the manufacturer/distributor, taking into account the age and condition of the endoscope, the reprocessing methods and methods of storage following last use.(for additional information refer to UK TSE Agents Safe working and the prevention of Infection Annex F Endoscopy guidance https://assets.publishing.service.gov.uk/government/uploads/system/uploads/attachment_data/file/470292/ACDP_TSE_Annex_F_Oct_2015.pdf)

Note: before an instrument is quarantined it should be first decontaminated to the required standard (https://www.hse.ie/eng/about/who/qid/nationalsafetyprogrammes/decontamination/decontamination-of-rimd.html) for additional information refer to Annex E Quarantining of surgical instruments guidance https://assets.publishing.service.gov.uk/government/uploads/system/uploads/attachment_data/file/547149/Annex_E_August_2016.pdf and Annex C - General principles of decontamination and waste disposal https://assets.publishing.service.gov.uk/government/uploads/system/uploads/attachment_data/file/427855/Annex_C_v3.0.pdf

NB: Single use biopsy forceps should be used in all patients

TABLE 3: PUBLIC HEALTH ACTIONS REQUIRED FOR ASYMPTOMATIC PATIENTS AT INCREASED RISK OF VCJD THROUGH:

- **Donating blood to someone who later developed vCJD**
- **Receiving blood from someone who has also given blood to a patient who went on to develop vCJD**
- **Surgery using instruments previously used on someone who developed vCJD**
- **Receiving blood from 300 or more UK donors since Jan 1990**
- **Treatment with certain UK sourced plasma products between 1990 and 2001 (inclusive)**

The following applies where an invasive procedure was carried out.

Tissue involved in the procedure	Action for instruments/RIMD is determined by the number of cycles of **use and decontamination** they have **already** been through since used on the index patient				Patients exposed to instruments/RIMD
	Action for surgical instruments/ RIMD by number of uses to date		Action for flexible endoscopes* by number of uses to date		
High infectivity • Brain • Spinal cord • Cranial nerves • Cranial ganglia • Posterior eye • Pituitary glands	Fewer than 20 uses Destroy or retain for exclusive use on this patient	More than 20 uses Reprocess & return to use	Fewer than 20 uses Destroy or retain for exclusive use on this patient	More than 20 uses Reprocess & return to use	No patients should traced and notified
Medium infectivity • Spinal ganglia • Olfactory epithelium*	Fewer than 10 uses Destroy or retain for exclusive use on this patient	More than 10 uses Reprocess & return to use	Fewer than 10 uses Destroy or retain for exclusive use on this patient	More than 10 uses Reprocess & return to use	No patients should traced and notified
Medium infectivity • Tonsil, adenoids • Appendix • Spleen • Thymus • Adrenal gland • Lymph nodes & gut-associated lymphoid tissues (including the rectum)	Fewer than 10 uses Destroy or retain for exclusive use on this patiente	More than 10 uses Reprocess & return to use	Fewer than 10 uses Reprocess & return to use **NB***	More than 10 uses Reprocess & return to use	No patients should traced and notified
Low infectivity All other tissues not listed above	Reprocess & return to use	Reprocess & return to use	Reprocess & return to use	Reprocess & return to use	No patients should traced and notified

Footnotes Table 3:

* Flexible endoscopes used on medium infectivity tissues may be returned to general used providing they have been decontaminated according to national standards (https://www.hse.ie/eng/about/who/qid/quality-and-patient-safety-documents/endoscope-reprocessing-version22.pdf) , with additional infection control precautions as described in the "Management and decontamination of flexible endoscopes (HTM 01-06)" https://www.gov.uk/government/publications/management-and-decontamination-of-flexible-endoscopes

Also:

TSE Agents Safe working and the prevention of Infection Annex F Endoscopy guidance https://assets.publishing.service.gov.uk/government/uploads/system/uploads/attachment_data/file/470292/ACDP_TSE_Annex_F_Oct_2015.pdf

**The olfactory epithelium is normally located deep within the nasal turbinates and normal nasal endoscope procedures do not reach the olfactory epithelium. However its distribution varies between individuals. 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 such contamination cannot be excluded, take precautions appropriate for medium infectivity tissues.

***NB Please note for surgical procedures involving lymphoid tissue, where endoscopes are used as well as instrumentation, assessment of the endoscope for contamination needs to be made. Annex M (general surgery) separates the risk between instruments that have been in direct contact with cut surfaces of medium infectivity tissues and those that haven't. https://assets.publishing.service.gov.uk/government/uploads/system/uploads/attachment_data/file/209780/Annex_M_-_Mananging_vCJD_risk.pdf

Note: before an instrument is quarantined it should be first decontaminated to the required standard (https://www.hse.ie/eng/about/who/qid/nationalsafetyprogrammes/decontamination/decontamination-of-rimd.html)

for additional information refer to Annex E Quarantining of surgical instruments guidance https://assets.publishing.service.gov.uk/government/uploads/system/uploads/attachment_data/file/547149/Annex_E_August_2016.pdf and Annex C - General principles of decontamination and waste disposal https://assets.publishing.service.gov.uk/government/uploads/system/uploads/attachment_data/file/427855/Annex_C_v3.0.pdf

NB: Single use biopsy forceps should be used in all patients

TABLE 4: PUBLIC HEALTH ACTIONS REQUIRED FOR: ASYMPTOMATIC PATIENTS AT INCREASED RISK OF IATROGENIC CJD (OTHER THAN VCJD) THROUGH:

The following applies where an invasive procedure has been carried out

- **Treatment with growth hormone from UK sourced human pituitary glands (before 1987)**
- **Treatment with gonadotropin derived from human pituitary glands for fertility treatment (before 1987)**
- **A neurosurgical procedure, or an operation for a tumour or cyst of the spine, before July 1996 on a patient who received (or might have received) a graft of human derived dura mater**
- **Surgery using instruments previously used on someone who developed CJD (other than vCJD)**

The following applies where an invasive procedure was carried out.

Tissue involved in procedure	Action for instruments/RIMD is determined by the number of cycles of **use and decontamination** they have **already** been through since used on the index patient				Patients exposed to instruments/RIMD
	Action for surgical instruments by number of uses to date		Action for flexible endoscopes* by number of uses to date		
High infectivity • Brain • Spinal cord • Cranial nerves • Cranial ganglia • Posterior eye • Pituitary glands	Fewer than 20 uses Destroy or retain for exclusive use on this patient	More than 20 uses Reprocess & return to use	Fewer than 20 uses Destroy or retain for exclusive use on this patient	More than 20 uses Reprocess & return to use	No patients should traced and notified
Medium infectivity • Spinal ganglia • Olfactory epithelium*	Fewer than 10 uses Destroy or retain for exclusive use on this patient	More than 10 uses Reprocess & return to use	Fewer than 10 uses Destroy or retain for exclusive use on this patient	More than 10 uses Reprocess & return to use	No patients should traced and notified
Low infectivity All other tissues not listed above	Reprocess & return to use	Reprocess & return to use	Reprocess & return to use**	Reprocess & return to use	No patients should traced and notified

*The olfactory epithelium is normally located deep within the nasal turbinates and normal nasal endoscope procedures do not reach the olfactory epithelium. However its distribution varies between individuals. 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 such contamination cannot be excluded, take precautions appropriate for medium infectivity tissues.

** Flexible endoscopes used on medium infectivity tissues may be returned to general used providing they have been decontaminated according to national standards (https://www.hse.ie/eng/about/who/qid/quality-and-patient-safety-documents/decontamination/hse-standards-and-recommended-practices-for-endoscope-reprocessing-units.html), with additional infection control precautions as described in the "Management and decontamination of flexible endoscopes (HTM 01-06)" https://www.gov.uk/government/publications/management-and-decontamination-of-flexible-endoscopes

Also:

TSE Agents Safe working and the prevention of Infection Annex F Endoscopy guidance https://assets.publishing.service.gov.uk/government/uploads/system/uploads/attachment_data/file/470292/ACDP_TSE_Annex_F_Oct_2015.pdf

Note: before an instrument is quarantined it should be first decontaminated to the required standard (https://www.hse.ie/eng/about/who/qid/nationalsafetyprogrammes/decontamination/decontamination-of-rimd.html) for additional information refer to Annex E Quarantining of surgical instruments guidance https://assets.publishing.service.gov.uk/government/uploads/system/uploads/attachment_data/file/547149/Annex_E_August_2016.pdf and Annex C - General principles of decontamination and waste disposal ttps://assets.publishing.service.gov.uk/government/uploads/system/uploads/attachment_data/file/427855/Annex_C_v3.0.pdf

NB: Single use biopsy forceps should be used in all patients

TABLE 5: PUBLIC HEALTH ACTIONS REQUIRED FOR SYMPTOMATIC PATIENTS WITH AN UNCLEAR DIAGNOSIS:

- **Diagnosis is unclear but not vCJD**
- **Diagnosis is unclear but vCJD is still being considered. A greater range of medium risk tissues should be considered during the risk assessment if variant CJD has not been ruled out as a potential diagnosis.**

Irreversible actions such as permanent disposal of instruments and notification of subsequently exposed patients should not be conducted until the diagnostic status is clear.

The following applies where an invasive procedure was carried out.

Tissue involved in procedure	Action for instruments/RIMD is determined by the number of cycles of **use and decontamination** they have **already** been through since used on the index patient				Patients exposed to instruments/RIMD
	Action for surgical instruments/ RIMD by number of uses to date		Action for flexible endoscopes* by number of uses to date		
High infectivity • Brain • Spinal cord • Cranial nerves • Cranial ganglia • Posterior eye • Pituitary glands	Fewer than 20 uses Remove from general use Quarantine pending diagnosis	More than 20 uses Reprocess & return to use	Fewer than 20 uses Remove from general use Quarantine pending diagnosis	More than 20 uses Remove from general use Quarantine pending diagnosis	No patients should traced and notified until the diagnosis is clear
Medium infectivity • Spinal ganglia • Olfactory epithelium* If variant CJD is still being considered: • Tonsil; appendix; spleen, adenoids • Thymus; adrenal gland • Lymph nodes & gut-associated lymphoid tissues (including rectum)	Fewer than 10 uses Remove from general use Quarantine pending diagnosis	More than 10 uses Reprocess & return to use	Fewer than 10 uses Remove from general use Quarantine pending diagnosis	More than 10 uses Remove from general use Quarantine pending diagnosis	No patients should traced and notified until the diagnosis is clear
Low infectivity All other tissues not listed above	Reprocess & return to use	Reprocess & return to use	Reprocess & return to use	Reprocess & return to use	No patients should traced and notified

* The olfactory epithelium is normally located deep within the nasal turbinates and normal nasal endoscope procedures do not reach the olfactory epithelium. However its distribution varies between individuals. 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 such contamination cannot be excluded, take precautions appropriate for medium infectivity tissues.

Also:

TSE Agents Safe working and the prevention of Infection Annex F Endoscopy guidance https://assets.publishing.service.gov.uk/government/uploads/system/uploads/attachment_data/file/470292/ACDP_TSE_Annex_F_Oct_2015.pdf

Note: before an instrument is quarantined it should be first decontaminated to the required standard (for additional information refer to TSE Agents Safe working and the prevention of Infection Annex E Quarantining of surgical instruments guidance https://assets.publishing.service.gov.uk/government/uploads/system/uploads/attachment_data/file/547149/Annex_E_August_2016.pdf).

NB: Single use biopsy forceps should be used in all patients

APPENDIX 6:
Algorithm on pre-operative assessments

Algorithm on pre-operative assessments

Before:
- intradural surgical procedures
- posterior ophthalmic surgical procedures
- intradural neuro-endoscopic procedures

Team member complete **pre-operative CJD/ vCJD risk assessment form** with patient

Overall patient care should remain the priority

If an emergency surgery event and patient unable to answer questions:
- Ask family member
- Ask GP

If no definitive answer: procedure occurs but **all instruments to be quarantined**

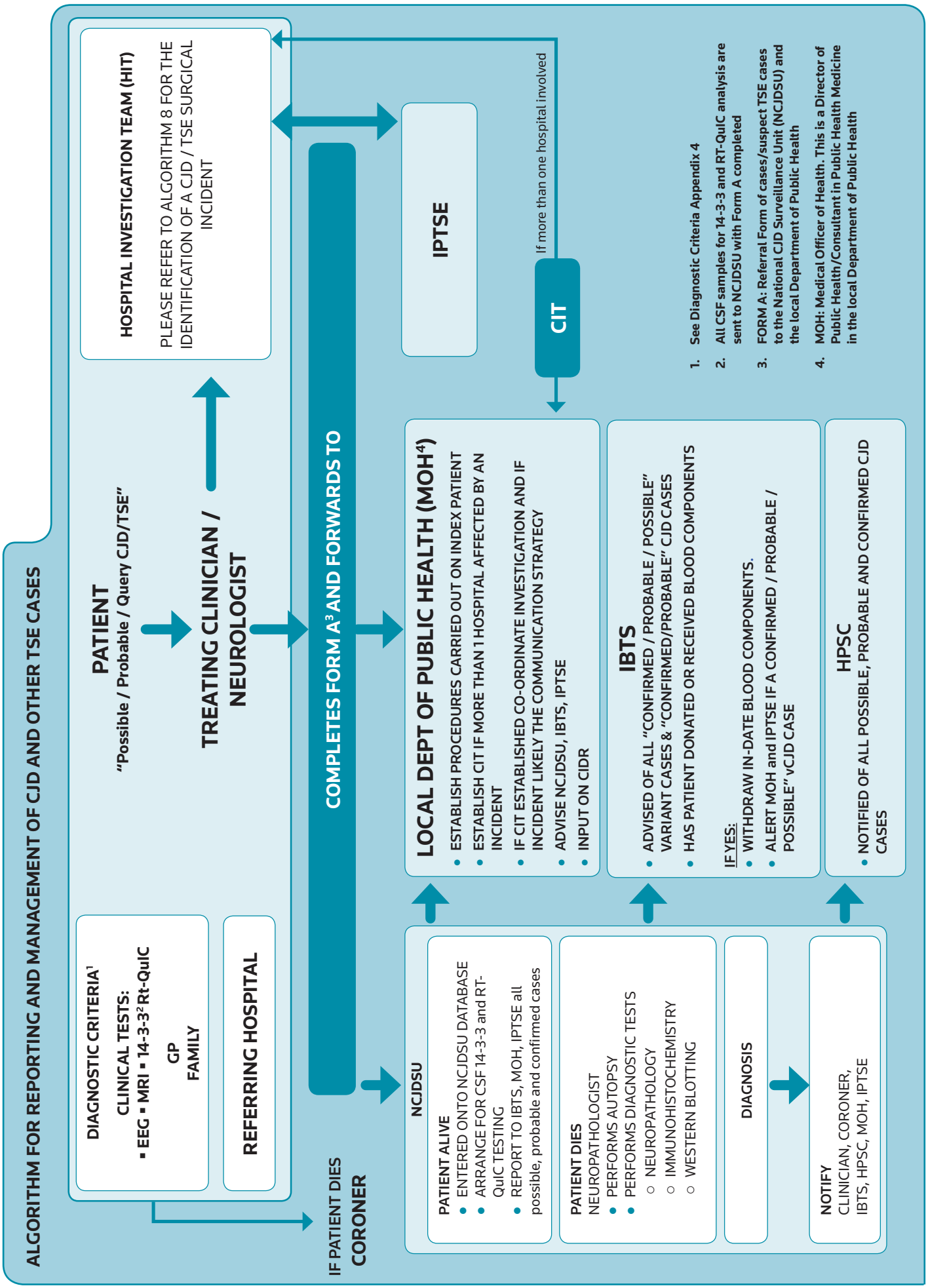
If the patient answered YES to any question on form:
- notify Infection Prevention and Control immediately AND prior to proceeding with the procedure
- Do further investigation into the patient's CJD/vCJD risk
- Consult with IPTSE - if not practical and CJD risk NOT outruled, please note guidance below.

If patient considered to be at increased risk for CJD/vCJD by the IPTSE OR CJD risk NOT outruled:

- Special infection prevention and control precautions should be taken for all surgery or endoscopy involving contact with medium or high infectivity tissues including quarantining of instruments
- Part 4 of the Public Health England guidance provides advice on the precautions during the treatment and Annex F provides information on endoscopic procedures

If patient considered to be at increased risk for CJD/vCJD by the IPTSE:
- ONLY then should the patient be informed
- This risk ascertainment should be recorded in patient's medical notes and communicated to their GP.

APPENDIX 7:


ALGORITHM FOR REPORTING AND MANAGEMENT OF CJD AND OTHER TSE CASES
DIAGNOSTIC CRITERIA[1]
CLINICAL TESTS:
▪ EEG ▪ MRI ▪ 14-3-3[2] Rt-QuIC
GP
FAMILY
REFERRING HOSPITAL
PATIENT
"Possible / Probable / Query CJD/TSE"
TREATING CLINICIAN / NEUROLOGIST
HOSPITAL INVESTIGATION TEAM (HIT)
PLEASE REFER TO ALGORITHM 8 FOR THE IDENTIFICATION OF A CJD / TSE SURGICAL INCIDENT
IPTSE
If more than one hospital involved
CIT
COMPLETES FORM A[3] AND FORWARDS TO
LOCAL DEPT OF PUBLIC HEALTH (MOH[4])
• ESTABLISH PROCEDURES CARRIED OUT ON INDEX PATIENT
• ESTABLISH CIT IF MORE THAN 1 HOSPITAL AFFECTED BY AN INCIDENT
• IF CIT ESTABLISHED CO-ORDINATE INVESTIGATION AND IF INCIDENT LIKELY THE COMMUNICATION STRATEGY
• ADVISE NCJDSU, IBTS, IPTSE
• INPUT ON CIDR
IBTS
• ADVISED OF ALL "CONFIRMED / PROBABLE / POSSIBLE" VARIANT CASES & "CONFIRMED/PROBABLE" CJD CASES
• HAS PATIENT DONATED OR RECEIVED BLOOD COMPONENTS
IF YES:
• WITHDRAW IN-DATE BLOOD COMPONENTS.
• ALERT MOH and IPTSE IF A CONFIRMED / PROBABLE / POSSIBLE" vCJD CASE
HPSC
• NOTIFIED OF ALL POSSIBLE, PROBABLE AND CONFIRMED CJD CASES
IF PATIENT DIES
CORONER
NCJDSU
PATIENT ALIVE
• ENTERED ONTO NCJDSU DATABASE
• ARRANGE FOR CSF 14-3-3 and RT-QuIC TESTING
• REPORT TO IBTS, MOH, IPTSE all possible, probable and confirmed cases
PATIENT DIES
NEUROPATHOLOGIST
• PERFORMS AUTOPSY
• PERFORMS DIAGNOSTIC TESTS
○ NEUROPATHOLOGY
○ IMMUNOHISTOCHEMISTRY
○ WESTERN BLOTTING
DIAGNOSIS
NOTIFY
CLINICIAN, CORONER, IBTS, HPSC, MOH, IPTSE
1. See Diagnostic Criteria Appendix 4
2. All CSF samples for 14-3-3 and RT-QuIC analysis are sent to NCJDSU with Form A completed
3. FORM A: Referral Form of cases/suspect TSE cases to the National CJD Surveillance Unit (NCJDSU) and the local Department of Public Health
4. MOH: Medical Officer of Health. This is a Director of Public Health/Consultant in Public Health Medicine in the local Department of Public Health

APPENDIX 8:

Algorithm for identification of a CJD/vCJD Incident*

Follow the flowchart below to assess surgical & endoscopic procedures that have been carried out on people with CJD or at increased risk of CJD

A symptomatic CJD case or suspected CJD case is identified

A patient is newly identified as at increased risk of CJD

An invasive procedure has been carried out on a patient with or at increased risk of CJD where TSE instrument precautions were not followed

Action: assess CJD type
Discuss with clinician (NCJDSU and IPTSE as required)
Action: Lookback - review past procedures- Dept Public Health & Hospital Incident Team*
Use GP and hospital records to review surgical and endoscopic procedures over the appropriate lookback period, depending on CJD type

SYMPTOMATIC CJD/vCJD PATIENT (CONFIRMED / POSSIBLE / PROBABLE / DIAGNOSIS UNCLEAR[1])

PATIENT AT INCREASED RISK OF CJD

IS IT SPORADIC/ INHERITED/ IATROGENIC OR TYPE OF CJD UNCERTAIN (NOT VCJD)	IS IT VCJD/VCJD (IATROGENIC) OR TYPE OF CJD UNCERTAIN (AND VCJD IS BEING CONSIDERED)	AT INCREASED RISK OF INHERITED[2] TSE?	BLOOD RECIPIENT (TRANSFUSION FROM DONOR WITH vCJD)?	OTHER AT INCREASED RISK OF CJD?
HIT / CIT REVIEW INVASIVE PROCEDURES WHILST SYMPTOMATIC AND 8 YEARS BEFORE[3]	HIT / CIT: REVIEW INVASIVE PROCEDURES SINCE 1980[4]	REVIEW INVASIVE PROCEDURES OVER PAST 8 YEARS[3]	REVIEW PROCEDURES SINCE TRANSFUSION	REVIEW PROCEDURES OVER PAST 12 MONTHS[5]

HAS A RELEVANT INVASIVE PROCEDURE BEEN IDENTIFIED IN THE RELEVANT TIME PERIOD?

IF **YES OR UNCLEAR**, THEN THIS MAY BE AN INCIDENT

IF **NO** THEN **NOT** AN INCIDENT

HOSPITAL INVESTIGATION TEAM (HIT)

- IDENTIFY INVASIVE PROCEDURES INVOLVING HIGH / MEDIUM RISK TISSUES[6]
- IDENTIFY RIMD USED DURING THE INVASIVE PROCEDURE
- LOCATE AND QUARANTINE RIMD/INSTRUMENTS – IPTSE WILL ADVISE FURTHER
- ESTABLISH IF OTHER PATIENTS EXPOSED
- COMPLETE FORM B[7]
- FORWARD FORM B TO THE LOCAL DEPT OF PUBLIC HEALTH (MOH)[9] AND IPTSE[8].

IPTSE (INCIDENT PANEL ON TRANSMISSIBLE SPONGIFORM ENCEPHALOPATHIES)

ADVISORY ROLE IN THE EVENT OF AN INCIDENT ON:

- TRANSMISSION PREVENTION
- RIMD/ INSTRUMENT QUARANTINE, RELEASE OR DESTROY
- COLLECTION OF PATIENT INFORMATION
- CONTACT TRACING
- NOTIFICATION OF EXPOSED PATIENTS IF REQUIRED
- COMMUNICATION STRATEGY IF REQUIRED
- OTHER MEASURES TO PROTECT THE WIDER PUBLIC

Notes:
*adapted from the Public Health England and Health Protection Scotland document *Public Health Action following a report of a new case of CJD or a person at increased risk of CJD*
https://assets.publishing.service.gov.uk/government/uploads/system/uploads/attachment_data/file/474338/CJD_public_health_action_new_case_301015.pdf

[1] Please see diagnostic criteria in Appendix 4 for guidance.

[2] Please refer to Appendix 3 of the document "Public Health Action following a report of a new case of CJD or a person at increased risk of CJD https://assets.publishing.service.gov.uk/government/uploads/system/uploads/attachment_data/file/474338/CJD_public_health_action_new_case_301015.pdf

[3] Infectivity in sporadic, inherited prion disease and iatrogenic CJD (apart from iatrogenic vCJD) is considered significant only in the last 8 years of the incubation period.

[4] As little is known about the development of tissue infectivity in humans infected with vCJD, please review surgical procedures since 1980, this is the date when first exposure to vCJD through the food chain is estimated.

[5] 12 months lookback will identify instruments that could potentially still pose a significant onward transmission risk. Notification of patients exposed in these incidents is not required.

[6] **High risk procedures:**
<u>All types of CJD:</u>
- Brain
- Spinal cord
- Cranial nerves (specifically the entire optic nerve and only the intracranial components of the other cranial nerves)
- Cranial ganglia
- Posterior eye (specifically the posterior hyaloid face, retina, retinal pigment epithelium, choroid, subretinal fluid, optic nerve)
- Pituitary gland

[6] **Medium risk procedures:**
<u>All types of CJD:</u>
- Spinal ganglia
- Olfactory epithelium

<u>vCJD:</u>
- Tonsil
- Lymph nodes and other organised lymphoid tissues containing follicular structures
- Gut-associated lymphoid tissue
- Appendix
- Adrenal gland
- Spleen
- Thymus

[7] Appendix 12: Form B CJD incident reporting form to the incident panel on transmissible spongiform encephalopathies (IPTSE) and the local Department of Public Health

[8] MOH: the Medical Officer of Health (MOH)/Director of Public Health (DPH) in the local department of public health. The Medical Officer of Health may be the Director of Public Health in the region concerned, or a Consultant in Public Health Medicine (CPHM).

APPENDIX 9:

Algorithm for management of suspect CJD patients after death

REFERRING HOSPITAL

SUSPECT CJD/vCJD PATIENT

PATIENT DIES

TREATING CLINICIAN

PRONOUNCES DEATH

MUST NOTIFY CORONER
MUST NOTIFY NCJDSU
01-809 2631 or if out of hours 01-809 3000 asking for neuropathologist on call.

IF CORONER DOES NOT DIRECT FOR AUTOPSY, CLINICIAN MAY REQUEST CONSENT FOR AUTOPSY FROM FAMILY.

INFORMS FAMILY:

- CORONERS CASE
- IF AUTOPSY GOES AHEAD, ORGANS RETAINED AFTER PM FOR ~2 MONTHS
- RETAINED ORGANS MAY BE:
  - RETURNED TO FAMILY

  OR
  - RESPECTFULLY DISPOSED OF BY BEAUMONT HOSPITAL

*SEE FORM C APPENDIX 13 AND FORM RELATING TO THE CREMATION OF BODY PARTS.[21]

CORONER

- IF DECIDES AUTOPSY WARRANTED, REQUESTS POST MORTEM IN WRITING-FAX TO BEAUMONT HOSPITAL (NEUROPATHOLOGY)

REFERRING HOSPITAL STAFF

- OBTAIN GARDA ID OF BODY AND GIVE THIS TO BODY TRANSPORTER
- ENSURES BODY IS TAGGED
- USE BODY BAG
- INCLUDE CHART WITH BODY
- MAY INCLUDE FORM C WITH BODY

REFERRING HOSPITAL MORTICIAN

- ORGANISE BODY TRANSPORT TO BEAUMONT HOSPITAL FOR PM
- INCLUDE CHART WITH BODY
- MAY INCLUDE FORM C WITH BODY

BEAUMONT HOSPITAL

AUTOPSY

BODY RETURNED ~48h
OPEN CASKET ALLOWED

BRAIN RETAINED
(~2 Months)

RESPECTFULLY DISPOSED OF BY BEAUMONT HOSPITAL

RETURNED TO FAMILY UNDERTAKER

21 Example of form relating to the cremation of body parts https://www.glasnevintrust.ie/funeral-services/downloads/

APPENDIX 10:
CONTACT DETAILS

A

National CJD Surveillance Unit (NCJDSU)
Department of Neuropathology, Beaumont Hospital,
Dublin 9
(01) 809 3000
Neuropathologist francesabrett@beaumont.ie
Neuropathologist michaelfarrell@beaumont.ie
Neuropathologist alanbeausang@beaumont.ie
Neuropathologist janecryan@beaumont.ie
Medical Scientist teresaloftus@beaumont.ie

B

Departments of Public Health (Medical Officers of Health)
For an updated list of the Medical Officers of Health please refer to the following website:
http://www.hpsc.ie/NotifiableDiseases/Whotonotify/File,13160,en.pdf

East (Counties Dublin, Kildare and Wicklow)
Medical Officer of Health,
Department of Public Health,
Room G29,
Dr Steevens' Hospital,
Dublin 8.
Phone: 01 6352145
Fax: 01 6352103

Midlands (Counties Laois, Offaly, Longford and Westmeath)
Medical Officer of Health,
Department of Public Health,
Area Office,
Arden Road,
Tullamore,
Co. Offaly.
Phone: 057 9359891
Fax: 057 9359907

Mid West (Counties Clare, Limerick and North Tipperary)
Medical Officer of Health,
Department of Public Health,
Mount Kennett House,
Henry Street,
Limerick.
Phone: 061 483337
Fax: 061 464205

North East (Counties Cavan, Louth, Meath and Monaghan)
Medical Officer of Health,
Department of Public Health,
Railway Street,
Navan,
Co. Meath.
Phone: 046 9076412
Fax: 046 9072325

North West (County Donegal)
Medical Officer of Health,
Department of Public Health,
Iona House,
Upper Main Street,
Ballyshannon, Co. Donegal.
Phone: 071 9852900
Fax: 071 9852901

Counties Sligo and Leitrim
Medical Officer of Health,
Department of Public Health,
Bridgewater House,
Rockwood Parade,
Sligo.
Phone: 071 9174750
Fax: 071 9138335

South (County Cork)
Medical Officer of Health,
Department of Public Health,
Floor 2,
Block 8,
St Finbarr's Hospital,
Douglas Road,
Cork.
Phone: 021 4927601
Fax: 021 4923257

County Kerry
Medical Officer of Health,
Department of Public Health,
Rathass,
Tralee,
Co. Kerry.
Phone: 066 7184548
Fax: 066 7184542

South East (Counties Carlow, Kilkenny, South Tipperary, Waterford and Wexford)
Medical Officer of Health,
Department of Public Health,
Lacken,
Dublin Road,
Kilkenny.
Phone: 056 7784142
Fax: 056 7784599

West (Counties Galway, Mayo and Roscommon)
Medical Officer of Health,
Department of Public Health,
Merlin Park Hospital,
Galway.
Phone: 091 775200
Fax: 091 758283

C

Irish Blood Transfusion Service (IBTS)
Dr Niamh O'Flaherty,
Irish Blood Transfusion Service,
National Blood Centre,
James Street,
Dublin 8.
Phone: (01) 4322868 / 800

D

Incident Panel on Transmissible Spongiform Encephalopathies (IPTSE)

Members:
Chair: Dr Kevin Kelleher,
Assistant National Director
Strategic Planning and Transformation
Public Health and Child Health,
Health Service Executive,
Mount Kennett House,
Henry Street,
Limerick
Tel: (061) 483 347
Fax: (061) 464 205
Email: kevin.kelleher@hse.ie

Dr Brian O'Connell
Consultant Microbiologist
St. James's Hospital

Brian O'Mahony
Chief Executive
Irish Haemophilia Society

Dr Colette Bonner
Deputy CMO
Department of Health

Dr Emer O'Connell
Consultant in Public Health Medicine
Department of Public Health, Galway

Dr Francesca Brett
Consultant Neuropathologist
Beaumont Hospital

Dr Robert Cunney
Consultant Microbiologist
Temple Street Children's University Hospital

Sabine Rowland
Sterivigilance Nurse
St. James's Hospital

Sheila Donlon
ADON Infection Prevention and Control
Cavan Monaghan Hospital

Dr Siobhan Hutchinson
Consultant Neurologist
St. James's Hospital

Carl O'Regan
CJD Surveillance Officer
Department of Neuropathology
Beaumont Hospital

Caroline Conneely
National Decontamination Quality Lead
Quality Improvement Division
Health Service Executive
Dr. Steevens Hospital,
Steevens Lane,
Dublin 8

E

Consultant Neurologist and member of the Incident Panel on Transmissible Spongiform Encephalopathies (IPTSE)
Dr Siobhán Hutchinson
Consultant Neurologist
Neurology Department
St. James's Hospital
Dublin 8
Tel: 01 410 3000

F

Health Protection Surveillance Centre (HPSC)
25-27 Middle Gardiner Street
Dublin 1
Tel: 01 8765 300
Fax: 01 8765 384
Web: www.hpsc.ie

G

Caroline Conneely
National Decontamination Quality Lead
Quality Improvement Division
Health Service Executive
Dr. Steevens Hospital
Steevens Lane
Dublin 8
E-mail: caroline.conneely1@hse.ie
Tel: 01 6352355

APPENDIX 11:

FORM A: Referral of cases/suspect TSE cases to the National CJD Surveillance Unit (NCJDSU)[1] and to the Local Department of Public Health (Also to be adapted with outcomes data)

Patient Details:

Patients Name:		Maiden Name:	
DOB:		Today's Date:	
Address:		GP Name & Address:	
Referring Consultant Name:			
Attending Hospital:		Assessed by Neurologist?	
Contact No.:		Neurologist details:	
If possible vCJD: donated/received blood/plasma components?		All types TSE: donated/received tissues/organs?	
MOH/Public Health notified?[2]		NCJDSU Notified?	

Clinical Details: Symptomatic No ☐ Yes ☐

Classification[3]:

☐ **Sporadic CJD**	☐ **Familial CJD**
☐ **Variant CJD**	☐ **Iatrogenic CJD**

☐ **Definite CJD**	☐ **Possible CJD**
☐ **Probable CJD**	☐ **Diagnosis Unclear[4]**

☐ **At increased risk of CJD**

Date of first symptoms?

Symptoms:
Any of the following:

☐ Myoclonus	☐ Ataxia
☐ Pyramidal features	☐ Cerebellar problems
☐ Extrapyramidal features	☐ Psychiatric symptoms
☐ Akinetic mutism	☐ Sensory symptoms
☐ Chorea / Dystonia	☐ Visual symptoms

EEG?	[Triphasic Periodic Discharge (1/sec)?]
Brain MRI?	[Caudate/putamen (sCJD) **OR** pulvinar (vCJD) high signal?]
CSF 14-3-3?	
Biopsies Performed?	

Diagnostic Outcome:

Confirmed CJD ☐
CJD thought unlikely ☐
Not CJD ☐

Details re diagnostic outcome:

HAS any of the following applicable CJD or vCJD incident occurred?: YES NO

- o A patient has donated organs/tissues before being diagnosed with CJD or vCJD
- o A patient has donated blood before being diagnosed with vCJD
- o A patient has donated organs/tissues before being identified as having an increased risk of CJD or vCJD
- o A patient has donated blood before being identified as having an increased risk of vCJD
- o A patient with confirmed/probable/possible diagnosis of CJD or vCJD has had an invasive procedure involving high or medium level risk tissues within the likely infective period and appropriate infection control guidance was not followed
- o A patient with an increased risk of CJD or vCJD had an invasive procedure involving high or medium level risk tissues and appropriate infection control guidance was not followed

CSF Specimen Details:

Date of Sampling:		**Date CSF sent:**	
Storage Conditions:	4°C ☐	-20°C ☐	-70°C ☐
White cell count		**Red cell count**	
CSF Total Protein			

The National CJD Surveillance Unit will not process CSF samples without receipt of this completed form.

[1]**NCJDSU:** National CJD Surveillance Unit
Department of Neuropathology
Beaumont Hospital
Dublin 9
Tel: (01) 809 2631

e-mail: francescabrett@beaumont.ie or michaelfarrell@beaumont.ie

[2]**MOH:** MOH is the Director of Public Health (DPH) or a designated Consultant in Public Health Medicine (CPHM). For relevant contact details please see http://www.hpsc.ie/NotifiableDiseases/Whotonotify/File,13160,en.pdf

[3] **Classification** For guidance refer to the Diagnostic Criteria and Case Definitions
http://www.hpsc.ie/a-z/other/cjd/casedefinitions/

Diagnosis unclear: some cases, especially early in the course of the disease may not reach the diagnostic criteria of possible CJD, but may still be suspected as cases of CJD.

APPENDIX 12:

Form B: CJD incident reporting form to the Incident Panel on Transmissible Spongiform Encephalopathies (IPTSE) and the local Department of Public Health

Guidance Note

Please use this form to report relevant surgical or other invasive procedures, to the Incident Panel on Transmissible Spongiform Encephalopathies (IPTSE) and the relevant Department of Public Health
All requests for advice from the Panel should be addressed to:

Dr Kevin Kelleher, Assistant National Director – Strategic Planning and Transformation – Public Health and Child Health
Health Service Executive, Mount Kennett House, Henry Street, Limerick
Tel: +353 (0) 61 483347
Fax: +353 (0) 61-464205
email: kevin.kelleher@hse.ie

Please ensure that this form is returned to us securely, either by encrypted email, safe fax or post.

To be completed/given by IPTSE	
PI number:	
To be completed by member of Hospital Investigation Team (usually clinician with primary responsibility)	
A. REPORTER'S CONTACT DETAILS	
Name:	Position:
Organisation:	
Address:	
Telephone number:	
E-mail:	
B. CLINICIAN WITH PRIMARY RESPONSIBILITY (IF DIFFERENT FROM REPORTING DOCTOR) CONTACT DETAILS	
Name:	Position:
Address:	
Telephone number:	
E-mail:	
B. DIRECTOR OF PUBLIC HEALTH/ CONSULTANT IN PUBLIC HEALTH MEDICINE (CPHM) CONTACT DETAILS	
Name:	Position:
Address:	
Telephone number:	
E-mail:	

B. INDEX PATIENT DETAILS			
1. Date of birth:		2. Age:	3. Sex:
4. Date of onset of symptoms:			
5. Date of first presentation to clinician:			
6. Alive?: Yes ☐ No ☐	If dead, date of death:		

CJD TYPE/DIAGNOSIS	**Please insert 'Yes' or 'No'**

7. For index patients with symptoms of CJD, please give details of the patient's diagnosis:-

	CJD diagnosis status at time of reporting			
Index patient CJD type/diagnosed	definite	probable	possible	not known
sporadic				
variant				
familial/genetic				
iatrogenic				
iatrogenic vCJD				
Other, please give details:				

9. Has the diagnosis been confirmed by the National CJD Surveillance Unit, (NCJDSU)? Yes ☐ No ☐ Not known ☐

NCJDSU case identification number:_____

If not confirmed, please give details:

_____

_____

Other comments:

8. Is the index patient 'at increased risk of CJD'? **This applies to both symptomatic and asymptomatic cases**	**Please insert 'Yes' or 'No'**
Did the patient have the following exposures:	
a) Has the index patient received a blood transfusion donated by someone who later developed vCJD?	
b) Has the index patient donated blood to someone who later developed vCJD?	
c) Has the index patient received a blood transfusion from a blood donor who also gave blood to someone who developed vCJD?	
d) Has the index patient received certain UK sourced plasma products (such as clotting factors) between 1990 and 2001?	
e) Was the index patient identified as having received blood or blood components from 300 or more UK donors since 1990?	
f) Is the index patient at risk of CJD for a different reason? (section 1) If yes, please give details.	

C. INVASIVE PROCEDURES WITH HIGH/MEDIUM INFECTIVITY TISSUES FOR CJD/vCJD *(a patient may have undergone one or several procedures. Please complete a new sheet for each procedure). Please complete all questions.*	***For IPTSE use*** Incident No: **Procedure No:**
1. When did the procedure take place?	
2. Hospital/other healthcare setting	Name: Address:
3. What procedure was carried out?	
4. What speciality was the procedure?	
5. What anaesthetic equipment was involved (if any)	
6. Did the procedure include endoscopy? If yes, please give details.	

7. Which tissues were involved? Please tick		Any notes/details?
☐	Brain	
☐	CNS / Spinal cord	
☐	Posterior eye	
☐	Olfactory epithelium	
☐	Tonsil / appendix / spleen / thymus	
☐	Other lymphoid tissue	
☐	Other (please give available details):	

8. How many reusable invasive medical devices (RIMD) including sets and supplementary single instruments /devices were used, and what were they? *(Please continue overleaf or attach details if necessary)*	
9. What type of decontamination procedures are used for these RIMD?	
10. Were standard decontamination methods used?	
11. Are there any reasons to suspect/doubt a problem with decontamination processes? Please specify.	
12. How many times have the RIMD instruments been used and decontaminated since this procedure?	

13. Can you track and trace the RIMD including single supplementary instruments/devices through the decontamination process to the service user? (e.g. all / some / none / disposable / don't know)	
14. Where are the instruments now?* (e.g. all are quarantined / some are quarantined / none are quarantined / not applicable/ don't know/ destroyed)	
15. How many people might have been exposed to the RIMD / instruments (or pool of instruments)?	
16. Did any staff member receive an exposure to blood and body substance during the invasive procedure or during the decontamination processes[22]?	
17. Other comments (*Please continue overleaf or attach if necessary)*	

* Following a report of a new case of CJD/person at increased risk, the hospital should ensure surgical instruments (including endoscopes) that have potentially been in contact with high or medium infectivity tissues for CJD, and have been through fewer than 10 cycles for medium risk tissue or 20 cycles for high risk tissue, are decontaminated as normal and removed from general use until the situation can be clearly risk assessed.

[22] For healthcare workers: Percutaneous or muco-cutaneous inoculation of tissues or blood from probable or confirmed cases of all types of human prion diseases including CJD. For laboratory workers: Percutaneous or muco-cutaneous inoculation of tissues or blood from TSE-infected animals or tissues. For more information please see page 14 in this link. https://assets.publishing.service.gov.uk/government/uploads/system/uploads/attachment_data/file/427854/Infection_controlv3.0.pdf

D.. HAS THE DONATION OF ORGAN/TISSUE/BLOOD COMPONENTS BEEN IDENTIFIED (**BLOOD COMPONENTS ONLY APPLICABLE IF A CASE OF VCJD):** YES☐NO☐	***For IPTSE use*** Incident No: **Procedure No:**

1. What type of donation was made?

Donation Of Organ

Donation Of Tissue

Donation Of Blood Components

2 If applicable which tissues/organ were involved?

Dates

_____

_____

_____

_____

_____

_____

3. If blood components applicable: Details of each blood component unit ID numbers are required and dates of transfusion

Date of transfusion		**Unit ID numbers**	**Details. Number of recipients etc**
Date			
Date			

4. Other comments (*Please continue overleaf or attach if necessary)*

Any notes/details?
Hospital/Other healthcare setting

E. Has the receipt of organ/tissue/blood components been identified? (**blood components are only applicable if a case of vCJD**): Yes ☐ No ☐	***For IPTSE use*** Incident No: **Procedure No:**

1. What type of procedure was carried out?

Receipt of organ

Receipt of tissue

Receipt of blood components

2. If applicable which tissues/organ were involved?	Any notes/details? Hospital/Other healthcare setting
Dates	

3. If blood components applicable: Details of each blood component unit ID numbers are required and dates of transfusion

Date of transfusion	**Unit ID numbers**	**Details**
Date		
Date		

APPENDIX 13:

Form C. Example of a CJD post mortem information form

Beaumont Hospital
Coroner Suspect CJD Autopsy Information Form

(This example form may be used when referring cases to the Coroner for CJD Post Mortem Examination)

This is not a consent form

The Coroner may order that a post mortem be performed on the body of _____
If this is the case, consent from the next of kin is not an option as the Coroner may, under the law, order a post mortem in certain situations to establish or clarify the cause of death. A post mortem involves the removal and detailed examination of the deceased brain. It is usual for the brain to be retained during the procedure and for small samples of tissue to be taken for microscopic examination.

PRE CORONER'S DECISION REGARDING POST MORTEM

I confirm that:

- I understand that the option for my consent does not arise for a Coroner's post mortem examination ☐
- I have been informed the reason why this death was reportable to the Coroner ☐
- I have been informed that tissue and organ will be retained following the post mortem for further diagnostic examination ☐

PLEASE READ THE OPTIONS GIVEN BELOW CAREFULLY

- I agree that a member of Beaumont Hospital Staff will contact me following the post mortem examination to provide advice and on-going information regarding organ retention. ☐

OR

- I do not want the hospital to make any further contact with me in relation to the organ retention. ☐
- In declining contact I am informed that the retained organs will be respectfully disposed of by Beaumont Hospital. ☐

Signed:_____ Date: DD / MM / YYYY Relation to deceased:_____
(Nominated family member)

Contact Number: (Mobile)_____(Home)_____

NAME (PLEASE PRINT):_____

Address:_____

_____

I confirm that I have explained the Coroner's Post Mortem procedure and the organ retention to the nominated family member.

Signed:_____ Date: DD / MM / YYYY Contact Number:_____
(Health Care Professional)

NAME (PLEASE PRINT):_____

APPENDIX 14:
Acknowledgements

The following groups contributed to the development of this document.

- Lead author: Dr. Emer O'Connell
- Current and past members of the Incident Panel on Transmissible Spongiform Encephalopathies (IPTSE) Dr Joan O'Riordan, Paschal Kent, Teresa Loftus and Joy Markey with special mention of Sabine Rowland and Dr Rachel Howley.
- A subgroup of the Public Health Medicine Communicable disease Group: Drs. Emer O'Connell (Chair), Áine McNamara, Margaret O'Sullivan, Mary Ward, Sinead Donohue, Rose Fitzgerald, Anthony Breslin and Peter Finnegan.
- Ms Kirsty MacKenzie, HPSC and Dr Katy Sinka, CJD Section Head, Public Health England.
- Submissions received from consultations (Appendix 15)

APPENDIX 15

Organisations consulted during consultation period

The consultation document was made available via the HPSC website, www.hpsc.ie, highlighted in Epi Insight and emailed directly to the following:

Academy of Clinical Science and Laboratory Medicine

An Bord Altranais

Association of Internal Medicine

Health and Safety Authority

HIQA

HSE Nursing and Midwifery Services

Infection Prevention Society

Intensive Care Society of Ireland

Irish Blood Transfusion Service

Irish College of General Practitioners

Irish College of Ophthalmologists

Irish Dental Association

Irish Institute of Clinical Neuroscience

Irish Institute of Trauma and Orthopaedic Surgery

Irish Patients Association

Irish Society of Clinical Microbiologists

Irish Society of Gastroenterology

Neuroscience Ireland

Royal College of Physicians of Ireland

Royal College of Surgeons in Ireland

Specialists in Public Health Medicine

Health Protection Surveillance Centre

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