Cheshire and Wirral Partnership NHS
NHS Foundation Trust

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Creuzfeld Jakob Disease (CJD) Policy

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Type of document	Policy
Target audience	All CWP staff
Document purpose	CJD is the main prion disease in humans. Currently there is no evidence to suggest that CJD or variant CJD is transmitted from person to person via close contact and that the main risk in healthcare settings is via invasive procedures. This policy aims to ensure that all Trust staff understand how CJD is transmitted and the precautions that are necessary when caring for an affected service user.

Approving meeting	Infection Prevention and Control Sub Committee	Date July-17
Implementation date	Sept-17 followed by an annual compliance review	

CWP documents to be read in conjunction with	
HR6	Trust-wide learning and development requirements including the training needs analysis (TNA)
IC7	Patient isolation policy
IC2	Hand decontamination policy and procedure
IC3	Standard (Universal) infection control precautions policy
GR30	Decontamination policy and disinfection policy
IC1	Trustwide infection control operational policy
IC12	Care of the deceased and the appropriate use of body bags
IC16	Policy for handling of linen and clothing
HS1	Waste Management Policy

Document change history	
What is different?	Minor changes and addition of flowchart
Appendices / electronic forms	N/A
What is the impact of change?	Minimal impact.

Training requirements	Yes - Training requirements for this policy are in accordance with the CWP Training Needs Analysis (TNA) with Education CWP.

Document consultation	
East locality	Who within this service have you spoken to
Wirral locality	Who within this service have you spoken to
West locality	Who within this service have you spoken to
Corporate services	Who within this service have you spoken to
External agencies	Who within this service have you spoken to

Financial resource implications	None

External references
1. Advisory Committee on Dangerous Pathogens (2013)Retrieved from website 21 February 2017: http//:www.hse.gov.uk/aboutus/meetings/committees/acdp/index.htm
2. The National Creutzfeldt-Jakob Surveillance Unit (2017) Retrieved from website 21 February 2017: http/:www.cjd.ed.ac.uk
3. Minimise Transmission Risk of CJD and vCJD in healthcare Settings (PHE 2012). Retrieved from website 21 February 2017: http//:www.gov.uk
4. Position Statement on vCJD (Nov 2015). Retrieved from website 21 February 2017: http//:www.transfusinguidelines.org/...vcjd
5. Wilson, J. (2006). Infection Control in Clinical Practice . 2nd edition. London. Balliere Tindall

Equality Impact Assessment (EIA) - Initial assessment	**Yes/No**	**Comments**
Does this document affect one group less or more favourably than another on the basis of:		
- Race	No	
- Ethnic origins (including gypsies and travellers)	No	
- Nationality	No	
- Gender	No	
- Culture	No	
- Religion or belief	No	
- Sexual orientation including lesbian, gay and bisexual people	No	
- Age	No	
- Disability - learning disabilities, physical disability, sensory impairment and mental health problems	No	
Is there any evidence that some groups are affected differently?	No	
If you have identified potential discrimination, are there any exceptions valid, legal and/or justifiable?		
N/A		
Is the impact of the document likely to be negative? - If so can the impact be avoided? - What alternatives are there to achieving the document without the impact? - Can we reduce the impact by taking different action?	No N/A N/A N/A	
Where an adverse or negative impact on equality group(s) has been identified during the initial screening process a full EIA assessment should be conducted. If you have identified a potential discriminatory impact of this procedural document, please refer it to the human resource department together with any suggestions as to the action required to avoid / reduce this impact. For advice in respect of answering the above questions, please contact the human resource department.		
Was a full impact assessment required?	No	
What is the level of impact?	Low	

Contents

Quick reference flowchart

Patient identified with suspected vCJD

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Notify IPC team on 01244 397700

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Clinician in charge of patient care must report case to National CJD Surveillance Unit, National prion Clinic and the Consultant for Communicable Disease (CCDC) at Public Health England (PHE).

Further information:
http://www.cjd.ed.ac.uk/surveillance

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There is no evidence of risk of occupational transmission of the disease to any healthcare worker in contact with affected patients or their blood, body fluids or tissue.

The precautions documented in the Trust's standard (Universal) infection control precautions policy are sufficient to protect against infection

1. Introduction

Transmissible Spongiform Encephalopathies (TSEs), sometimes known as prion diseases are fatal degenerative brain diseases which occur in humans and certain other animal species. A common feature of all TSEs is the appearance of microscopic vacuoles in the grey matter of the brain, giving a sponge-like appearance. There are several recognised TSEs including Creutzfeldt Jakob Disease (CJD) in humans, Bovine Spongiform Encephalopathy (BSE) in cattle and Scrapie in sheep. Other TSEs include Gerstmann-Straussler-Scheinker Syndrome (GSS) which is an extremely rare form of TSE, Familial Fatal Insomnia and Kuru.

For the purpose of this policy the focus will remain on the CJD and VCJD as these TSEs are the ones most likely to be encountered within the Trust, however, it must be emphasised that these diseases remain extremely rare.

CJD and vCJD is caused by an abnormal disease-specific isoform of prion protein. The incubation period is unknown, possibly 15 months to over 20 years. CJD is a protracted, progressive illness of the CNS, characterised by progressive dementia or progressive unsteadiness and clumsiness – CJD, vCJD and all other TSEs are invariably fatal once clinical signs appear and there is no known treatment or prophylaxis.

TSEs agents exhibit an unusual resistance to conventional chemical and physical decontamination methods, which therefore presents a major challenge in the healthcare setting when dealing with contaminated items of equipment such as surgical instruments. They are not significantly affected by disinfectants, for example formalin and ethylene oxide and infectivity remains after autoclaving at conventional times and extreme temperatures, for example 121 degrees centigrade for 15 minutes.

2. Different types of CJD

2.1 Sporadic CJD

The majority of cases of classical CJD occur in the over 55s. This form of CJD is found worldwide with an incidence of one case per million population, per year. The disease tends to present with a clear neurological illness that progress rapidly. The cause of sporadic CJD remains uncertain - one theory suggests that the normal prion protein in the brain undergoes a spontaneous change to the abnormal form causing disease.

2.2 New variant CJD (now just called variant CJD or VCJD)

First reported in 1996, investigations have failed to provide any conclusive evidence to suggest that it is related to diet, as sporadic CJD has been reported in countries where BSE is not present. Unlike classical CJD, its initial presentation can be psychiatric or behavioural problems and it may not be clear that the individual has a neurological illness until several months later. Variant CJD tends to affect the younger age group with an average age of onset of illness of 27.

2.3 Genetic CJD

This is a very rare illness caused by an inherited abnormal gene. There is therefore no relationship between this form and BSE. In the majority of cases the illness is known within the family because of the family history. The definitive test to confirm the presence of CJD is a blood test.

2.4 Iatrogenic CJD

This is also extremely rare and is a CJD which has been accidentally transmitted during the course of medical or surgical procedures. For example, within the United Kingdom CJD was transmitted via human growth hormone treatment administered during childhood. The diagnosis can often be reached from previous medical or surgical treatment.

2.5 Diagnosis of CJD or VCJD

An absolute definitive diagnosis of any form of CJD requires neuropathological examination of brain tissue. Such a procedure is usually undertaken only during a post mortem examination – it is rare for

such an investigation to be carried out on a live person and is not usually necessary in the investigation of a possible case of CJD.
When a diagnosis of CJD is suspected investigations can be undertaken for the following reasons:

- To exclude other possible diagnoses;
- To support a CJD diagnosis.

The information described below has all been obtained from The National Creutzfeldt-Jakob Surveillance Unit and was correct at the time this policy was written.

2.6 Sporadic CJD

- **The EEG** – in sporadic CJD the normal electrical rhythms of the EEG are slowly lost;
- **Cerebrospinal fluid (CSF) 14-3-3 analysis** – whilst the routine examination of the CSF of patients with sporadic CJD is unremarkable the analysis for certain brain proteins, particularly 14-3-3 may be useful in diagnosis. 14-3-3 is a normal neuronal protein and can be released into the CSF in response to a variety of neuronal insults. It is therefore generally a non-specific finding and cannot be used as screening test as the following illnesses may also give a positive result:
    - Herpes simplex encephalitis and other viral encephalitis;
    - Recent cerebral infarction or haemorrhage;
    - Hypoxic brain damage;
    - Glioblastoma;
    - Carcinomatous meningitis;
    - Paraneoplastic encephalopathy.

However, if the above conditions can be excluded then a positive result, in an appropriate clinical context, is strongly supportive of a diagnosis of sporadic CJD.

Magnetic Resonance Imaging (MRI) – Computerised Tomography (CT) of the brain is usually normal in cases of sporadic CJD, although atrophy may be seen if the illness has progressed for some time. MRI can be used to exclude other illnesses and abnormalities may be seen in the anterior basal ganglia and cortex. Whilst such findings are currently not accepted into the clinical diagnostic criteria they may still be helpful in individual cases.

2.7 Variant CJD

MRI - Should be undertaken in all suspected cases of vCJD - at least to exclude any other underlying causes of illness. There is a characteristic abnormality seen in the posterior thalamic region which is highly sensitive and specific for variant CJD. Such a finding would indicate that a case of "possible variant CJD" is now a "probable variant CJD"

Tonsil biopsy – unlike other forms of CJD, vCJD has been identified in tissues such as the lymph nodes, spleen, tonsil and appendix. Therefore it is possible to find the disease related protein in a biopsy of such tissue. However, this does involve a surgical procedure with carries a risk of infectivity. Such biopsies are most useful in providing support for the diagnosis when other investigations such as MRI have proved inconclusive.

CSF 14-3-3 – a general CSF examination will be unremarkable. The 14-3-3 test as described previously is not as sensitive in variant CJD as it is in sporadic CJD. A positive result may provide support to the diagnosis; however, a negative result does not exclude the diagnosis.

2.8 Genetic CJD

The definitive test is analysis of a specific gene for appropriate mutations and can be performed on a simple blood sample. A family history of the disease may not always be present.

2.9 Iatrogenic CJD

A diagnosis can usually be made after obtaining an accurate history of relevant know risk factors such as receiving cadaveric human growth hormone or receiving a human dura mater graft in surgery.

2.10 Transmission of CJD via blood transfusions

There have been four cases of vCJD infection associated with blood transfusions with all four cases having received the transfusions between 1996 and 1999. The possibility that vCJD can be transmitted through blood raises concern about the possible infectivity of blood components and plasma products. However since April 2004 blood donations are no longer accepted from individuals who have received blood during the course of any medical procedure since January 1st 1980. In addition to this advice, since 1997, the National Blood Service has:

- Recalled any blood products from donors who later go on to develop vCJD;
- Imported plasma from countries with few or no cases of BSE to manufacture blood products;
- Removed white blood cells from the blood before it is transfused as such cells may carry vCJD infection;
- Imported plasma from countries with few or no cases of BSE for children born after 1996.

2.11 Risk assessment

There is no evidence of occupational transmission of the disease to medical, nursing or laboratory staff in contact with affected patients or their blood / body fluid / tissue. Therefore the precautions documented in the Trust's standard (Universal) infection control precautions policy are sufficient to protect against infection.

3. Categorisation of patient/service user groups at risk of CJD or vCJD

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

A. Symptomatic patients

- Patients / service users who fulfil the diagnostic criteria for definite, probable or possible CJD or vCJD;
- Patients / service users with neurological disease of unknown aetiology who do not fit the criteria for possible CJD or vCJD, but where the diagnosis of CJD is being actively considered.

B. Asymptomatic patients / service users at risk from familial forms of CJD linked to genetic mutations

- Individuals who have or have had two or more blood relatives affected by CJD or other prion disease, or a relative known to have a genetic mutation indicative of familial CJD;
- Individuals who have been shown by specific genetic testing to be at significant risk of developing CJD or other prion disease.

C. Asymptomatic patients/service users potentially at risk from iatrogenic exposure

- Recipients of hormone derived from human pituitary glands, e.g. growth hormone, gonadotrophin;
- Individuals who have received a graft of dura mater. Individuals who underwent neurosurgical procedures or operations for a tumour or cyst of the spine before August 1992 may have received a graft of dura mater and should therefore be treated as "at risk" unless evidence demonstrates that dura mater was not used;
- Patients/service users who have been contacted as potentially "at risk" because of exposures to instruments used on, or receipt of blood, plasma derivatives, organs or tissues donated by a donor who subsequently went on to develop CJD or vCJD.
- Individuals with bleeding disorders i.e. congenital and acquired haemophilia (Haemophilia A and Haemophilia B), Von Willebrand Disease, other congenital bleeding disorders and congenital antithrombin III deficiency, who have been treated with UK-sourced pooled factor concentrates or antithrombin (i.e. clotting factors and antithrombin made from pooled plasma. These include factor VIII, factor IX, factor VII, factor XI, factor XII and prothrombrin complex concentrates as well as antithrombin between 1980 and 2001are classified as at risk of vCJD for public health purposes.

4. Infection prevention and control precautions

The Infection Prevention and Control Nurses (IPCNs) must be notified of any service users suspected as being at risk of CJD or vCJD. The clinician leading on a service user's care must inform the IPCNs as soon as possible if they suspect a service user has CJD. The clinician in charge of the service user's care must report a suspected case of CJD to the National CJD Surveillance Unit, the National Prion Clinic and the local Consultant in Communicable Disease Control (CCDC). Further information and the appropriate documentation can be found at: http://www.cjd.ed.ac.uk/surveillance

Any occurrence of suspected infection will be reported to Public Health England (PHE) by the clinician in charge of the service user's care.

It is not necessary to isolate the service user in a side room. Patients with CJD or vCJD can be cared for in an open ward using Standard Precautions – please refer to the Trust's standard (Universal) infection control precautions policy for more information. For guidance on handling the service users clothing and linen please refer to the CWP policy for handling of linen and clothing policy.

If is it necessary to carry out any procedures on the service users where contamination with CSF or blood may occur, for example, lumbar puncture, biopsies or venepuncture then the advice in the Trust's standard (Universal) infection control precautions policy must be strictly adhered to.

All equipment used that may come into contact with blood or CSF must be **single use only** and must be disposed of as infectious waste immediately after use. All other waste, including Personal Protective Equipment (PPE) must be place in an infectious waste bag and incinerated as per the CWP waste management policy. Any sharps used during the procedure must be disposed of immediately as per the CWP standard (Universal) infection control precautions policy. A **SENIOR** member of staff in the pathology department must be informed **BEFORE** the specimens are sent.

Any blood / body fluid spillages that occur must be dealt with promptly as per the CWP standard (Universal) infection control precautions policy. It is imperative that any investigations such as venepuncture are carried out in area that will tolerate hypochlorite solution being used as this is the product recommended for cleaning up spillages of blood / body fluid.

If the member of staff carrying out the procedure sustains an injury, such as a sharps injury then the prevention and management of exposure to health care associated infections (HCAI) and inoculation incidents policy must be implemented and Occupational health are to be informed at the earliest opportunity.

Such procedures must only be carried out if deemed clinically necessary and by experienced and appropriately trained staff only.

4.1 Surgical interventions

As CWP does not carry out any surgical procedures, this topic will not be discussed any further in this policy.

5. Accident reporting and health surveillance

A CWP incident form must be completed detailing all accidents and occurrences with infectious or potentially infectious material involving the exposure of individuals. This applies whether or not the accident was reportable under The Reporting of Incidents, Diseases and Dangerous Occurrences Regulations, 1995 (RIDDOR). CWP staff must ensure that they attend occupational health as soon as possible after any incident where they may have been exposed to infectious or potentially infectious material.

Employers are required to maintain a confidential list of employees exposed to the CJD agent in the following circumstances:

- There is a deliberate intention to work with the agent in cases of accidental exposure when a local risk assessment in conjunction with the IPCNs shows there is a significant risk of exposure.

This record will involve recording any known incident or accident with blood, CSF or suspect tissues and sharps injuries / splashes. This list must be kept for forty years after last known exposure on the staff member's occupational health record and it must indicate the type of work done.

6. Care of the deceased

Please refer to the Trust's Policy for the care of the deceased and the appropriate use of body bags for further information.