

# Transmissible Spongiform Encephalopathy (TSE) including Creutzfeldt- Jacob Disease (CJD) Variant CJD (vCJD)

This policy describes the process for managing patients with suspected with TSE including CJD and vCJD within Leicestershire Partnership NHS Trust.

|   |  |                                       |
|---|--|---------------------------------------|
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## Version Control and Summary of Changes

| <b>Version</b> | <b>Date</b>   | <b>Comments<br/>(description change and amendments)</b>   |
|----------------|---------------|---|
| Version 1      | May 2014      | <p>Review of national guidelines for policy relevant to LPT.</p> <p>Infection Prevention and Control advice pertaining to Theatres, day surgery and endoscopy departments is currently provided by UHL infection prevention and control team. Therefore staff are directed to this team for advice pertaining to these services.</p> <p>Circulated for comments to relevant parties within LPT and outside organisations.</p> |
| Version 2      | July 2017     | <p>Clarification that no new national guidelines available.</p> <p>Policy updated in line with the latest LPT template for policies.</p> <p>Information relating to UHL services removed as not relevant to LPT.</p>  |
| Version 3      | November 2019 | <p>Clarification that no new guidelines available.</p> <p>Policy updated in line with the latest LPT template for policies</p>  |
| Version 4      | December 2020 | <p>Clarification that no new guidelines available</p> <p>Policy updated in line with the latest LPT template for policies</p>   |

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## Definitions that apply to this policy

|  |  |
|--|--|
| <b>Transmissible Spongiform Encephalopathy (TSE)</b> | TSE is a group of diseases that are caused by the build-up of an abnormal form of the naturally occurring prion protein in the brain   |
| <b>Creutzfeldt-Jacob Disease (CJD)</b>               | A form of TSE. It is a rare and ultimately fatal degenerative disease  |
| <b>vCJD</b>  | Variant Creutzfeldt-Jacob disease (vCJD) is a <a href="#">prion disease</a> that was first described in 1996 in the United Kingdom. There is now strong scientific evidence that the agent responsible for the outbreak of prion disease in cows, <a href="#">bovine spongiform encephalopathy</a> (BSE or 'mad cow' disease), is the same agent responsible for the outbreak of vCJD in humans. |
| <b>Classical CJD</b>                                 | Classical CJD is a human prion disease. It is a neurodegenerative disorder with characteristic clinical and diagnostic features. This disease is rapidly progressive and always fatal. Infection with this disease leads to death usually within 1 year of onset of illness.   |
| <b>Healthcare Acquired Infection (HCAI)</b>          | Infections that are acquired as a result of healthcare interventions.  |
| <b>Iatrogenic</b>                                    | Brought about by surgical or medical treatment.  |
| <b>Infection</b>                                     | Invasion of the body by organisms causing disease  |
| <b>Prion</b>   | A small infectious particle composed of abnormally folded protein that causes progressive neurodegenerative conditions. These abnormally folded proteins do not multiply in the host organism that they infect. Instead they affect the brain structure by acting as a template, inducing proteins with normal folding to convert to the abnormal prion form.                                    |
| <b>Standard Precautions</b>                          | The precautions taken by all staff for all patients all of the time based on a risk  |

## **1.0 Purpose of the policy**

The aim of this policy is to give direction to staff within LPT with regards to caring for patients with suspected Transmissible Spongiform Encephalopathy (TSE), including Creutzfeldt-Jacob Disease (CJD) and Variant Creutzfeldt-Jacob Disease (vCJD).

The transmission risks and diagnosis of the disease are identified, including the necessary infection prevention and control precautions with the overall aim being to reduce the risk of transmission.

The policy sets out to ensure that that all staff employed by LPT provide evidence based care which is in accordance with the Health & Social Care Act (2008), (updated 2015).

## **2.0 Summary and scope of policy**

There is currently no known cure for TSE, CJD or vCJD, nor is there currently any formal means of diagnosis except following the death of the patient. Therefore it is imperative that staff are aware of signs and symptoms of the disease and of the necessary precautions they need to take when TSE, CJD or vCJD is suspected.

Patients with TSE, CJD or vCJD are not an infection risk to other patients or staff and therefore will not require source isolation precautions.

This disease is thought to be caused by the build-up of an abnormal form of prion proteins that are found in the brain; and can be transferred by medical equipment following certain procedures which are deemed to be high risk. None of these 'high risk' procedures are currently undertaken within LPT services.

The emergence of the new viral infection – Covid-19 has not had an impact on the measures to be taken with a patient who is known to have TSC, CJD or vCJD.

## **3.0 Introduction.**

The purpose of this policy is to provide information with regards to the transmission of TSE, CJD and vCJD and to enable staff to ensure that they are following the correct procedures when caring for patients with suspected TSE, CJD or vCJD.

TSE is a group of diseases that affects both humans and animals. It is thought to be caused by the build-up of an abnormal form of the naturally occurring 'prion' protein in the brain. CJD is a form of TSE and is a rare and ultimately fatal degenerative disease.

CJD can be classed as classical or sporadic. It can also be genetic or a form of inherited prion disease and these are associated with mutation in the prion protein gene. Rarer forms of human prion disease included acquired diseases such as iatrogenic CJD. Iatrogenic CJD is very rare and occurs when CJD is transmitted as a result of medical or surgical exposures. To reduce this risk precautionary measures have been taken to improve the standards of surgical instruments and endoscope decontamination.

vCJD is thought to be most likely to be acquired through ingestion of meat contaminated with a bovine spongiform encephalopathy agent.

#### **4.0 The management of patients with Transmissible Spongiform Encephalopathy (TSE) including Creutzfeldt-Jacob Disease (CJD) Variant CJD (vCJD)**

This policy covers all healthcare settings within LPT.

#### **4.1 Diagnosis**

Diagnosis is usually made on a clinical basis; there are currently no widely available laboratory tests for human TSEs. At the present time diagnosis can only be confirmed by examination of brain tissue after death (post mortem). Brain biopsy may be used in investigating cases of suspected TSE but may not be definitive in establishing a diagnosis.

#### **4.2 Treatment**

There are currently no cures available, any treatment given would be to alleviate any symptoms if possible.

#### **4.3 Screening**

There is currently no screening process available that can give a conclusive diagnosis.

#### **4.4 Infection prevention and control measures**

A patient with suspected TSE is not an infection risk, therefore any patient who has been clinically 'provisionally' diagnosed with TSE does not require source isolation precautions. Standard precautions, such that are applicable to all patients at all times should be undertaken.

- Effective hand hygiene before and after each patient contact and contact with their environment is of paramount importance in reducing spread of all infections. Please refer to the LPT hand hygiene policy.
- Antibiotics must be prescribed according to the antibiotic guidance for primary care.
- Personal protective equipment must be worn where there is a risk of exposure to blood or body fluids. Please refer to the LPT personal protective policy.
- Cleaning and decontamination of equipment must be undertaken as per the LPT cleaning and decontamination policy.

There have been 4 cases of presumed person-to-person transmission of vCJD infection via blood transfusion reported within the United Kingdom since 2003 (3 clinical and 1 asymptomatic) and a further probable vCJD infection via plasma products has been reported in a haemophiliac person.

Since 1997, when the theoretical risk of vCJD transmission through blood was first considered, the UK blood services have taken a number of precautionary measures to protect the blood supply and associated plasma products. These include:

- Blood components, plasma products or tissues obtained from any person who later develops vCJD are withdrawn or recalled to prevent their use.
- Since 1998 plasma for the manufacture of plasma products, such as clotting factors, has been obtained from non-UK sources.
- Since 1998 synthetic (recombinant) clotting factor for treatment of haemophilia has been provided to those under the age of 16 and for all patients in whom it is suitable since 2005.
- Since 1999 white blood cells (which may carry a significant risk of transmitting vCJD) have been reduced in all blood used for transfusion. This process is known as leucodepletion.
- Since 2002 fresh frozen plasma for treating babies and young children born on or after 1 January 1996 has been obtained from the USA. In 2005 its use was extended to all children up to the age of 16.
- Since 2004, individuals who have received a transfusion of blood components since January 1980, or are unsure if they have a blood transfusion, are excluded from donating blood or platelets.
- Since 2009 cryoprecipitate, a special cold-treated plasma preparation, has been imported from the USA for children up to the age of 16

Even though the risk of obtaining CJD through blood and bodily fluids is very low and samples from patients with or suspected of having or at increased risk of CJD should be treated as potentially infectious and any samples sent to the laboratory should indicate that the patient from whom the sample is taken is suspected of having TSE, CJD or vCJD should this be the case.

When patients suspected of TSE, CJD or vCJD have certain procedures undertaken in theatres, including day surgery and endoscopy then the equipment used may need to be quarantined until a diagnosis can be made, and ultimately disposed of if a positive diagnosis is made. However LPT do not currently support these services and therefore this is not pertinent to LPT.

#### **4.5 Movement and transport of patients**

There are no infection prevention and control requirements to be implemented, other than standard precautions that are required for all patients, when patients are being moved from one area to another or transported in vehicles, including volunteer cars, private cars or ambulance transportation.

#### **4.6 Caring for patients within the community setting**

People should not be dissuaded from routine contact with patients with TSE, CJD or vCJD as they are not thought to present a risk through normal social or routine clinical contact. No special measures over and above standard infection prevention and control precautions are generally required for caring for patients with TSE, CJD

or vCJD patients in the community, as it is unlikely that procedures will be adopted that will lead to contact with high or medium risk.

#### 4.7 Deceased patients

After the patient has died, contact the mortuary for up to date protocols. The deceased patient must be placed into a body bag prior to transporting to the mortuary and normal procedures for bodies where there is a known infection risk must be followed. Embalming should be avoided in confirmed or suspected cases.

In all cases, discuss with a Consultant Histopathologist, regarding post mortem examination. Post mortems on diagnosed or suspected TSE patients are to be carried out at Queen’s Medical Centre Nottingham.

#### 5.0 Training

All clinical staff must undertake Trust Infection Prevention and Control mandatory training bi-annually. Additional training on TSE will be provided by the Infection Prevention and Control Team if required.

#### 6.0 Monitoring Compliance and Effectiveness

| REFERENCE  | MINIMUM REQUIREMENTS                        | EVIDENCE FOR SELF ASSESSMENT     | PROCESS FOR MONITORING    | RESPONSIBLE INDIVIDUAL/GROUP | FREQUENCY OF MONITORING                                   |
|--|---|----------------------------------|---------------------------|------------------------------|---|
| Transmissible Spongiform Encephalopathy (TSE) including Creutzfeldt-Jacob Disease (CJD) Variant CJD (vCJD) | Health and Social Care Act<br>CQC Outcome 8 | Incidents, complaints and trends | Activity training reports | IPCG                         | As part of annual report or in view of a serious incident |



## 7.0 References and bibliography

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