Nottinghamshire Hospice
adding life to days

STANDARD OPERATING PROCEDURE

SOP016	
Subject	Management of Patients with Confirmed, Suspected or at Risk of having Creutzfeldt - Jakob Disease (CJD) or any Transmissible Spongiform Encephalopathy (TSE). (This policy is subject to periodic review and will be amended according to service development needs)
Applicable to	This policy applies to all staff, volunteers and contractors who work for or provide care on behalf of Nottinghamshire Hospice
Date issued	Aug 2021
Next review date	Aug 2026
Lead responsible for Policy	Director of Care
Policy Reviewed by	Infection Prevention and Control Team Nottingham CityCare Partnership Care Service Team
Notified to	Quality and Safety Group
Authorised by	Board of Trustees
Links to other Policies	Infection Prevention and Control Policy
Summary	This document aims to provide a clear understanding of Nottinghamshire Hospices Infection Control Policy.
Target Audience	The policy aimed at all staff, volunteers and contractors who work for or provide care on behalf of Nottinghamshire Hospice.

IMPORTANT NOTICE: Staff should always refer to the website or folder on the 'N' drive for the most up to date information. If the review date of this policy or procedure has expired staff should seek advice from their clinical lead or manager regarding the appropriate action to be taken.

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1. Abbreviations, Acronyms and Definitions

Transmissible Spongiform Encephalopathy (TSE): Transmissible spongiform encephalopathies (TSEs) are a family of diseases of humans and animals characterized by spongy degeneration of the brain with severe and fatal neurological signs and symptoms (WHO,2021).

Creutzfeldt - Jakob Disease (CJD): This is a TSE disease which is a rare, but fatal condition that affects the brain. It causes brain damage that worsens rapidly over time.

Variant Creutzfeldt–Jakob disease (vCJD). First described in 1996, Variant CJD (vCJD) is similar to CJD as it too is a TSE, yet there are notable differences. First, young people are affected, with an average age of death under 30 years. Second, the disease has a relatively longer duration of illness. Finally, it is strongly linked to exposure, probably through food, to BSE. Other human TSEs have not been linked to food exposure. The route of transmission of vCJD is not yet fully proven but it is generally accepted that it is transmitted through exposure to food contaminated by BSE (WHO, 2021).

Bovine Spongiform Encephalopathy (BSE): A disease which affect the central nervous system of cattle. **NOT** to be related to Creutzfeldt–Jakob disease in human

Prion protein: Naturally occurring cellular protein the function of which as yet remains unclear. It is thought they may play a role in the transport of messages between specific brain cells.

Prion diseases: A group of progressive neurodegenerative conditions; CJD and TSE are prion diseases. These exist in animals and humans.

2. Purpose

The Health and Social Care Act (DH 2008) stipulates that all providers of NHS services have procedures and policies in place to protect patients, workers and others who may be at risk of acquiring a Health Care Associated Infection (HCAI) including CJD. The purpose of ths SOP is to:

- Assist clinicians to care for people living with a prion disease of Creutzfeldt - Jakob Disease (CJD) or any Transmissible Spongiform Encephalopathy (TSE)
- Provide information underpinning safe working practice to reduce the risks of transmission of TSE agents, including CJD and vCJD from affected patients to other patients or staff within the primary care setting.
- Ensure compliance with the Health and Social Care Act (2008).

Creutzfeldt and Jakob first described the illness in the 1920's later known as Creuzfeldt – Jakob disease (CJD) and various forms have been identified since.

CJD falls into four categories: Sporadic, Variant, Familial and Iatrogenic.

- Sporadic: The commonest form of CJD arises spontaneously for no known reason. This is the cause of 85% of all CJD cases (WHO, 2020.

- Variant CJD (vCJD), first recognized in the United Kingdom in 1996 has been linked to bovine spongiform encephalopathy, (BSE).

- Familial CJD (fCJD) is an inherited condition and cases represent 20% to 15% of the total number of CJD cases. Gerstmann-Sträussler-Scheinker disease (GSS) and fatal familial insomnia (FFI) are very rare forms of CJD.

- Iatrogenic CJD infection is inadvertently transmitted usually from a case with sCJD in the course of medical/surgical treatment, e.g. human pituitary hormone therapy, human dura mater grafts, corneal grafts or neurological instruments.

(WHO,2020)

3. Evidence base and interaction with other policies and procedures

- Nottinghamshire Hospice Decontamination of Non-Invasive Medical Equipment and the Environment (SOP014)
- Nottinghamshire Hospice Hand Hygiene Policy (CS015)
- Nottinghamshire Hospice Personal Protective Equipment Policy for Infection Prevention and Control (CS020)
- Nottinghamshire Hospice Waste Management Policy (CS023)
- Nottinghamshire Hospice Safe Sharps & Blood Borne Virus Policy (CS021)
- Nottinghamshire Hospice Blood, Bodily Fluids & Vaccine Spillages (SOP011)
- Collection of Specimens (excluding blood specimens) (SOP013)
- www.nhs.uk (NHS, 2020)
- Recommended standards and strategies for surveillance, prevention and control of communicable diseases (WHO,1999)

4.0 https://www.who.int/zoonoses/diseases/variantcjd/en/ (WHO,2021)

Scope and responsibilities

This SOP has been developed for all staff working within Nottinghamshire Hospice caring for patients with confirmed, suspected or at risk of having Creutzfeldt - Jakob Disease or any Transmissible Spongiform Encephalopathy.

Chief Executive & Executive Team is responsible for:

- Ensuring that there are arrangements in place within the Organisation to support infection prevention and control, in particular the necessary policies and training to reduce the risk of infections being transmitted.

Infection Prevention & Control Team are responsible for:

- Updating this Standard Operating Procedure every five years or more regularly if required.
- Ensuring infection prevention and control training is available for Hospice Staff.
- The follow up of all patients in the community who have confirmed, suspected or at risk of having Creutzfeldt - Jakob Disease or any Transmissible Spongiform Encephalopathy.

CityCare Staff are responsible for:

- Reading this Standard Operating Procedure and ensuring that they are up to date with infection prevention and control training which will incorporate the principles for the precautions and management of CJD patients.
- Liaising with the patient, family, carers and care agencies to ensure they are aware of the individual management plan and how to access further information if required.

4. Equipment required

- Liquid soap and paper towel or Community Hand Hygiene Pack (if necessary)
- Personal Protective Equipment, chosen by risk assessment.
- Sharps Container (if necessary)
- Waste receptacle (if necessary)
- Spill Pack (if necessary

5. Procedure

Patients are categorised according to their risk. A distinction is made between symptomatic patients who fulfil the diagnostic criteria for CJD and those at an increased risk of developing CJD because of family or medical history:

Symptomatic patients	• Patients who fulfil the diagnostic criteria for definite, probable or possible CJD or vCJD. • Patients with neurological disease of unknown aetiology, who do not fit the criteria for possible CJD or vCJD, but where the diagnosis of CJD is being actively considered.
Patients "at increased risk" from genetic forms of CJD	• Individuals who have been shown by specific genetic testing to be at significant risk of developing CJD. • Individuals who have a blood relative known to have a genetic mutation indicative of genetic CJD. • Individuals who have or have had two or more blood relatives affected by CJD or other prion disease.
Patients identified as "at increased risk" of vCJD through receipt of blood from a donor who later developed vCJD	• Individuals who have received labile blood components (whole blood, red cells, white cells or platelets) from a donor who later went on to develop vCJD
Patients identified as "at increased risk" of CJD/vCJD through iatrogenic exposures	• Recipients of hormone derived from human pituitary glands, *e.g*. growth hormone, gonadotrophin, are "at increased risk" of transmission of sporadic CJD. In the UK the use of human-derived gonadotrophin was discontinued in 1973, and use of cadaver-derived human growth hormone was banned in 1985. However, use of human-derived products may have continued in other countries after these dates. • Individuals who underwent intradural brain or intradural spinal surgery before August 1992 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). • Individuals who have had surgery using instruments that had been used on someone who went on to develop CJD/vCJD or was "at increased risk" of CJD/vCJD. • Individuals who have received an organ or tissue from a donor infected with CJD/vCJD or at "increased risk" of CJD/vCJD. • Individuals who have been identified as having received blood or blood components from 300 or more donors since January 1990. • Individuals who have given blood to someone who went on to develop vCJD • Individuals who have received blood from someone who has also given blood to a patient who went on to develop vCJD. • Individuals who have been treated with certain implicated UK sourced plasma products between 1990 and 2001.

The following is a brief summary of the Infection Prevention and Control principles with reference to the Organisations relevant policy which gives more in-depth information.

Step	Action	Rationale / outcome	Risk management / additional direction
1.	Hand Hygiene	The single most important measure to reduce the spread of infections. Therefore it is essential that staff carry out effective hand decontamination before and after care activities with patients and after any activity or contact that contaminates the hands.	• Further information is available in the Hospice Hand Hygiene Policy available on the website in the 'N' drive.
2.	Protective Clothing which includes: • Disposable plastic apron or gown. • Latex gloves or a suitable alternative e.g. nitrile. • Mask, eye protection or a visor if splashing is likely to occur.	Whenever there is the risk of contact with blood or bodily fluids protective clothing should be worn regardless of the infection status of the patient. Use of protective clothing relies on the individual carrying out a risk assessment of the procedure/ care activity being undertaken and to ensure this is documented.	• Further information is available in the Hospice Personal Protective Equipment Policy for infection prevention and Control available on the website in the 'N' drive.
3.	Blood and Spillage	Careful attention to standard infection prevention and control precautions will minimise any risks from blood. Drug administration by injection should involve the same precautions used for all work of this type with any patient i.e. avoidance of sharps injuries and the safe disposal of sharps.	• A sharps injury or contamination of abrasions with blood or bodily fluid should be treated according to the following guidance: • Hospice Blood, Body Fluids and Vaccine Spillages Standard Operating Procedure on the website on the website in the 'N' drive. Hospice Safe Sharps and Exposure to Blood Borne Viruses Policy on the website on the 'N' drive

Step	Action	Rationale / outcome	Risk management / additional direction
4.	Clinical Waste	High or medium risk tissues from patients with, or “at increased risk” of, CJD or vCJD, should be incinerated. However, as identified in the primary care setting, waste will be from low risk tissues or body fluids and should follow normal management processes (DH 2010 part 4).	• For further information please refer to the Hospice Waste Management Policy available on the intranet on the website on the ‘N’ drive
5.	Bed Linen/ Clothing	Clothing can be washed as normal, although in the interests of general hygiene, soiled linen can be washed separately. In the hospice setting red alginate bags should be available for soiled linen. In the home environment, soiled linen should be washed as soon as possible on the hottest setting the fabric can tolerate.	• For further information please refer to the Hospice Uniform and Dress Code Policy available on the intranet on the website on the ‘N’ drive
6.	Care After Death	Last Offices remain the same; there are no restrictions to relatives or friends viewing or having contact with the deceased.	• A cadaver bag is required for transportation of the deceased to the mortuary / funeral directors. This should be labelled as ‘High Risk or Danger of Infection’ prior to transportation to the mortuary, in line with normal procedures for deceased patients with a known infection risk (DH, 2010).