NICE National Institute for Health and Care Excellence

NICE guidance

Reducing the risk of transmission of Creutzfeldt–Jakob disease (CJD) from surgical instruments used for interventional procedures on high-risk tissues

HealthTech guidance
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Your responsibility

This guidance represents the view of NICE, arrived at after careful consideration of the evidence available. When exercising their judgement, healthcare professionals are expected to take this guidance fully into account, and specifically any special arrangements relating to the introduction of new interventional procedures. The guidance does not override the individual responsibility of healthcare professionals to make decisions appropriate to the circumstances of the individual patient, in consultation with the patient and/or guardian or carer.

All problems (adverse events) related to a medicine or medical device used for treatment or in a procedure should be reported to the Medicines and Healthcare products Regulatory Agency using the Yellow Card Scheme.

Commissioners and/or providers have a responsibility to implement the guidance, in their local context, in light of their duties to have due regard to the need to eliminate unlawful discrimination, advance equality of opportunity, and foster good relations. Nothing in this guidance should be interpreted in a way that would be inconsistent with compliance with those duties. Providers should ensure that governance structures are in place to review, authorise and monitor the introduction of new devices and procedures.

Commissioners and providers have a responsibility to promote an environmentally sustainable health and care system and should assess and reduce the environmental impact of implementing NICE recommendations wherever possible.

Contents

This guidance replaces IPG196 and IPG666.

1 Recommendations

This guidance is for interventional procedures on tissues considered at high risk of transmitting Creutzfeldt–Jakob disease (CJD). These procedures on high-risk tissues are intradural surgery on the brain (including the pituitary gland) and spinal cord, neuroendoscopy, and surgery on the retina or optic nerve (see appendix D for a complete list).

Note that the abbreviation 'CJD' is used for both sporadic and variant CJD (vCJD), including surgically transmitted CJD (stCJD), unless otherwise specified.

The recommendations do not apply to any interventional procedures done on patients already known to have or thought to be at increased risk of CJD as previously defined by the Advisory Committee on Dangerous Pathogens' Transmissible Spongiform Encephalopathies subgroup. For these patients and patients in whom the risk of CJD cannot be ascertained by questioning them, or when a diagnosis of CJD cannot be excluded, the Advisory Committee on Dangerous Pathogens' guidance on transmissible spongiform encephalopathy agents: safe working and the prevention of infection must be followed.

Decontamination

1.1 All surgical instruments that come into contact with high-risk tissues during an interventional procedure must be kept moist and separated from other instruments until they are cleaned, and then disinfected and sterilised (decontaminated). This improves the efficacy of the decontamination process and is highly cost effective.

Set integrity and tracking

1.2 Surgical instruments that come into contact with high-risk tissues must not be

moved from one set to another and must remain within their individual sets. Maintaining set integrity reduces the risks associated with instrument migration (including infection) and makes it easier to trace instruments back to the patients they were used on.

Supplementary instruments

1.3 Supplementary instruments that come into contact with high-risk tissues must remain within the individual set to which they have been introduced. Supplementary instruments are those that are not part of a specific instrument set. If supplementary instruments are used with different sets, this would compromise set traceability and increases the risks associated with instrument migration.

Neuroendoscopy

1.4 Rigid neuroendoscopes (rather than flexible neuroendoscopes) should be used if possible. They should be of a type that can be steam sterilised and must be thoroughly cleaned and steam sterilised after each use.

Single-use instruments

1.5 The evidence on cost effectiveness does not support using sets of single-use instruments to reduce the risk of Creutzfeldt–Jakob disease (CJD) transmission.

Systems specifically for people born after 1996

1.6 The evidence on cost effectiveness does not support introducing systems to maintain separate sets of neuroendoscopes and reusable surgical instruments for use on high-risk tissues for people born after 1996.

1.7 Removing the requirement to use different instruments on high-risk tissues for people born after 1996 would not markedly increase the risk of surgical transmission of CJD.

Other relevant guidance

1.8 This guidance should be used with:

- the Advisory Committee on Dangerous Pathogens' Transmissible Spongiform Encephalopathies subgroup's previous guidance on transmissible spongiform encephalopathy agents: safe working and the prevention of Infection
- the Department of Health and Social Care's Health Technical Memorandum (HTM) 01-01: decontamination of surgical instruments (2016) and corresponding guidance in the devolved administrations' areas.

Further research

1.9 NICE may update this guidance after 3 years or sooner if important new information becomes available, including evidence on:

- the epidemiology of CJD, including data on the prevalence of CJD and its infectivity in the UK population
- the transmission of CJD by surgical instruments, including cases of CJD in which surgery is a possible route of transmission
- the cost effectiveness of single-use instruments for use in interventional procedures on high-risk tissues
- commercially available decontamination methods that are safe and cost effective against prions
- the systems for, and cost effectiveness of, maintaining set integrity and traceability of instruments.

2 Indication

The condition

2.1 Creutzfeldt–Jakob disease (CJD) is a progressive, fatal neurological disease affecting the brain. It is caused by pathological accumulation of a transmissible form of protein called a prion. CJD belongs to a wider group of neurodegenerative disorders known as transmissible spongiform encephalopathies (TSEs) that affect both humans and animals. People with CJD typically present with rapidly progressive dementia, usually accompanied by myoclonus and cerebellar ataxia. Most people die within 4 months of disease onset, in a mute and immobile state.

Epidemiology of CJD

2.2 The incidence of any type of CJD (based on published surveillance studies) is 1 to 2 cases per million of the population worldwide. There are 4 aetiological CJD categories:

- Sporadic CJD (sCJD) accounts for 85% to 90% of cases worldwide. The aetiology is not known. It has an annual incidence of 1 to 2 deaths per million of population per year. The general rate of age-adjusted detection of sCJD is increasing in the UK. Reasons for this include improved case ascertainment and an ageing population (in which there is a higher incidence).
- Inherited (genetic or familial) CJD accounts for 5% to 15% of cases or about 10 deaths in the UK per year. It is associated with pathogenic mutations in the prion protein gene.
- Variant CJD (vCJD) is a novel form of human prion disease, first recognised in the UK in 1996. It is believed to result from consumption of food derived from cattle infected with bovine spongiform encephalopathy (BSE), a fatal neurodegenerative disease that causes sponge-like changes in the brain. vCJD is characterised by extensive lymphoreticular tissue involvement and a

young age at onset (the mean age at death is 28 years, compared with 66 years for sCJD). The clinical course of vCJD is distinct from that of sCJD. People with vCJD frequently present with sensory and psychiatric symptoms that are uncommon in people with sCJD. They develop progressive neurological signs such as gait disturbance, ataxia and tremor. The median duration of illness is longer than for sCJD (14 months compared with 4 months). By 2016 there had been 178 cases of vCJD in the UK. Three cases are considered to have occurred through blood transfusion and 175 cases were related to dietary exposure to BSE. The prevalence of non-clinical vCJD (abnormal prion accumulation in tissues without clinical symptoms) in the general UK population is estimated to be 240 per million, based on retrospective analyses of appendix specimens. In the UK, between 1988 and 1996, a series of measures were put in place to reduce the risk of people being exposed to BSE. Over the past 8 years there have been 0 or 1 deaths per year in the UK attributed to vCJD.

- Iatrogenic CJD (iCJD) accounts for less than 1% of cases each year. It is the transmission of prions through surgical or medical procedures (especially from tissues with the highest concentration of prions, such as brain and posterior eye tissue) or human-derived products (growth hormone, gonadotropin, dura mater grafts and packed red blood cells). Surgically transmitted CJD (stCJD) is theoretically possible through prion-contaminated instruments that have been previously used on patients with CJD. This includes patients who are asymptomatic but infectious because neural tissue has a high infectious load, and there are difficulties in eradicating prions from surgical instruments. The most common causes of iCJD are historic use of human growth hormone and dura mater grafts, according to a review of worldwide iCJD cases published in 2012. In the UK, 85 iCJD cases were identified between 1970 and 2016. Eight were from dura mater grafts, 1 was from human gonadotrophin and 76 were from human growth hormone. There were 4 cases of possible stCJD through contaminated neurosurgical instruments between 1952 and 1974; 3 in the UK and 1 in France. The University of Sheffield's School of Health and Related Research (ScHARR) systematic review indicates that the risk of stCJD is currently low and no cases were reported between 2005 and 2018. However, there is uncertainty about the future risk of stCJD because of the potentially long incubation period of CJD, difficulties in eradicating prions from surgical instruments, the

presumed subclinical prevalence in the general population, and high levels of infectivity in the brain. Neurosurgical instruments used on people who are possibly carriers of CJD are handled in accordance with the Advisory Committee on Dangerous Pathogens' Transmissible Spongiform Encephalopathies risk management subgroup's guidance on safe working and the prevention of infection.

- The National CJD Research and Surveillance Unit report the number of deaths per year in the UK attributed to the 4 categories of CJD.

Incubation periods

2.3 Evidence from retrospective data in the ScHARR systematic review shows that the incubation period of iatrogenic CJD ranges from 1 to 42 years with durations towards the shorter end of the range reported in cases of stCJD. Incubation times might be affected by the recipient's genotype and the infecting prion strain or subtype of CJD.

Infectivity

2.4 The infectivity of CJD is likely to be moderated by a number of factors including the recipient's genotype, the infecting prion strain, and the route of transmission. There are limited data about infectious dose or infectious titre in humans. The ID_{50} is the dose that would give the person receiving it a 50% chance of becoming infected. High values are expressed in log or factor-of-10 terms. For example, 1 g of brain tissue can have an ID_{50} of 10^8 (8 log). This means it carries a dose of 100,000,000 ID_{50}. The model assumes that intracranial transfer of 0.01 micrograms of such brain tissue would result in the recipient having a 50% chance of becoming infected with CJD.

3 Committee considerations

Issues considered in this guidance

3.1 The following issues of clinical and decontamination practice were considered in terms of clinical and cost effectiveness, patient safety and the extent to which they reduce the risk of Creutzfeldt–Jakob disease (CJD) transmission:

- use of reusable and single-use instruments in surgical procedures
- arrangements for cleaning, sterilising and tracking of reusable surgical instruments.

Issues not considered in this guidance

3.2 The following issues were not considered in this guidance:

- Transfusion of blood or blood products, including occupational exposure to blood or body fluids. Several organisations provide advice on measures to reduce the risks from blood transfusion and exposure to blood in the workplace, including the Department of Health and Social Care Advisory Committee on the Safety of Blood, Tissues and Organs (SABTO), and the Advisory Committee on Dangerous Pathogens' Transmissible Spongiform Encephalopathies subgroup.
- Extracorporeal life-support machinery, including cardiopulmonary bypass, haemodialysis and ventilator equipment.
- The risk of CJD and variant CJD (vCJD) transmission through drugs, which is regulated by the Medicines and Healthcare products Regulatory Agency (MHRA).
- The safety of transplant grafts, which is the responsibility of the Department of Health and Social Care and SABTO.
- The decontamination and reuse of single-use instruments, for which the

MHRA has issued guidance advising against the reuse of such items.

- General dentistry.

The evidence

3.3 For detailed information on the evidence see the University of Sheffield's School of Health and Related Research (ScHARR) systematic review, which consists of the following evidence:

- Direct evidence is limited because of the rare nature of CJD. Eight systematic reviews on the incidence and prevalence of CJD in the general population in the UK, risk of transmission through surgery, incubation periods, infectivity, efficacy of decontamination procedures, adherence to NICE guidance by keeping surgical instrument sets together, complications from single-use instruments and the likelihood of patients who have had high-risk surgery returning for further surgery. The systematic reviews use historical case reports of surgically transmitted CJD (stCJD), observational data, case-control studies and animal studies. They are mostly descriptive, and no formal critical appraisal of study quality was done.
- An updated version of the model used in NICE's previous appraisal of patient safety and reduction of risk of transmission of CJD (IPG196) to assess the cost effectiveness of potential strategies to reduce the risk of stCJD: the updated model assumes that all genotypes are susceptible to stCJD infection and that patients with stCJD can be misdiagnosed with an alternative neurodegenerative disease. A calibration of predicted model inputs was done with the number of possible stCJD cases observed between 2005 and 2018. All assumptions and the calibration were agreed with the CJD advisory subcommittee. A formal elicitation exercise with members of the CJD advisory subcommittee was used to update the parameters used in the modelling.

3.4 Organisations representing patients provided submissions and representation at the CJD advisory subcommittee meetings.

4 Committee comments

4.1 The committee emphasised the importance of continued surveillance for all forms of Creutzfeldt–Jakob disease (CJD) to identify trends in incidence rates. It noted that there are effective systems for doing this in the UK.

4.2 The committee noted that the economic modelling suggests that keeping surgical instruments moist is the most cost-effective strategy, because it saves money and potentially increases societal health. Additional strategies aimed at reducing the future risk of surgically transmitted CJD (stCJD) do not appear to be cost effective.

4.3 The committee supported the need, as outlined in the Department of Health and Social Care's Health Technical Memorandum (HTM) 01-01: decontamination of surgical instruments (2016), for systems to be developed that allow both individual instruments and sets to be tracked and traced. This would avoid instrument migration.

4.4 The committee encouraged further research into the development of cost-effective decontamination methods that remove or destroy prions from instruments.

4.5 The committee noted that single-use instruments are the only way of completely avoiding the potential for the transmission of CJD infection by surgical instruments.

4.6 The committee emphasised that clinicians should comply with the Department of Health and Social Care's Health Technical Memorandum (HTM) 01-01: decontamination of surgical instruments (2016) and corresponding guidance in the devolved administrations' areas and other relevant guidance and standards.

4.7 The committee emphasised that this guidance applies to procedures on high-risk tissues and not to people at risk of CJD as defined in the Advisory Committee on Dangerous Pathogens' guidance on transmissible spongiform encephalopathy agents: safe working and the prevention of infection.

Appendix A: Creutzfeldt–Jakob disease advisory subcommittee members and NICE project team

Creutzfeldt–Jakob disease advisory subcommittee members

The subcommittee met 5 times between 2017 and 2018.

Mr Tom Cadoux-Hudson
Consultant neurosurgeon, John Radcliffe Hospital, Oxford

Dr Chris Carroll
Reader in systematic review and evidence synthesis, School of Health and Related Research, University of Sheffield

Dr Tom Clutton-Brock
Chair, NICE interventional procedures advisory committee; consultant in anaesthesia

Dr Andrew Cook
Vice chair, NICE interventional procedures advisory committee; consultant in public health medicine

Mr Nicholas Haden
Consultant neurosurgeon, Derriford Hospital, Plymouth

Dr David Hilton
Consultant neuropathologist, University Hospitals Plymouth NHS Trust

Mr Alistair Jenkins
Consultant neurosurgeon, Royal Victoria Infirmary, Newcastle

Dr Neil McGuire
Clinical director of devices, Medicines and Healthcare products Regulatory Agency

Professor Simon Mead
Consultant neurologist and clinical lead of the UK National Prion Clinic based at the National Hospital for Neurology and Neurosurgery

Professor Graham Medley
Professor of infectious disease modelling, London School of Hygiene and Tropical Medicine

Dr Katy Sinka
Consultant scientist and epidemiologist, Public Health England

Professor Colin Smith
Professor of neuropathology, University of Edinburgh

Mr John Thorne
Consultant neurosurgeon, Royal Manchester Children's Hospital and Salford Royal

Dr Jimmy Walker
Scientific leader in water microbiology and decontamination, Public Health England

Mr Barrie White
Consultant neurosurgeon, Nottingham University Hospitals

Mrs Rosemary Harris
Lay representative and Interventional Procedures Advisory Committee member

Mr Lister Firkin
Lay representative and Interventional Procedures Advisory Committee member (attended only first subcommittee meeting in June 2017)

Mrs Gillian Turner
National CJD coordinator, CJD Support Network (joined in September 2017 after first subcommittee meeting in June 2017)

NICE project team

Professor Kevin Harris
Programme director and clinical advisor, interventional procedures programme

Professor John Powell
Consultant clinical advisor, interventional procedures programme

Mrs Lakshmi Mandava
Technical analyst

Mrs Bijal Joshi
Programme manager

Ms Hawra Gulal and Miss Deonee Stanislaus
Coordinators

Mr Azad Hussain
Administrator

Appendix B: Contributors

These organisations provided specialist advice and comments:

- CJD Support Network
- Child Growth Foundation

Appendix C: Related advisory groups

A number of advisory committees, expert groups and academic units are addressing issues related to Creutzfeldt–Jakob disease (CJD) including developing the scientific basis of our understanding of the disease, improving decontamination practices across the NHS and minimising the risk of transmission. NICE has made every effort to coordinate with these groups to ensure that this guidance takes account of, and builds on, their work. Some members of the CJD advisory subcommittee are also members of other CJD committees, working groups and academic units. NICE has been represented on some of these groups.

The following is a list of some of the organisations in the UK that either fund surveillance, study or provide advice on the public health aspect of CJD:

- Department of Health and Social Care
- Medical Research Council Prion Unit at University College London Hospitals' National Prion Clinic
- University of Edinburgh National CJD Research & Surveillance Unit
- Public Health England
- Health Protection Scotland

Appendix D: High-risk procedures

The OPCS-4 codes for intradural operations on the brain and spinal cord are shown below.

Neurosurgery

This list may not be comprehensive.

All purposeful intradural brain and spinal cord surgery are regarded as high-risk and this guidance applies wherever there is contact with or possible contamination by brain or spinal cord tissue.

Extradural surgery, even in the presence of a cerebrospinal fluid leak, is not high risk and this guidance does not apply.

- A01 Major excision of tissue of brain
- A02 Excision of lesion of tissue of brain
- A03 Stereotactic ablation of tissue of brain
- A04 Open biopsy of lesion of tissue of brain
- A05 Drainage of lesion of tissue of brain
- A07 Other open operations on tissue of brain
- A08 Other biopsy of lesion of tissue of brain
- A09 Neurostimulation of brain
- A10 Other operations on tissue of brain
- A12 Creation of connection from ventricle of brain
- A13 Attention to component of connection from ventricle of brain
- A14 Other operation on connection from ventricle of brain
- A16 Other open operations on ventricle of brain

- A20 Other operations on ventricle of brain
- A22 Operations on subarachnoid space of brain
- A24 Graft to cranial nerve
- A25 Intracranial transection of cranial nerve
- A26 Other intracranial destruction of cranial nerve
- A29 Excision of lesion of cranial nerve
- A30 Repair of cranial nerve
- A31 Intracranial stereotactic release of cranial nerve
- A32 Other decompression of cranial nerve
- A33 Neurostimulation of cranial nerve
- A34 Exploration of cranial nerve
- A36 Other operations on cranial nerve
- A38 Extirpation of lesion of meninges of brain
- A39 Repair of dura
- A42 Other operations on meninges of brain
- B01 Excision of pituitary gland
- B02 Destruction of pituitary gland
- B04 Other operations on pituitary gland
- B06 Operations on the pineal gland
- L33 Operations on aneurysm of cerebral artery
- L34 Other open operations on cerebral artery
- A44.1 Chordectomy of spinal cord
- A44.2 Extirpation of lesion of spinal cord NEC

- A44.3 Excision of lesion of intradural intramedullary spinal cord
- A44.5 Excision of lesion of intradural extramedullary spinal cord
- A44.8 Other specified partial extirpation of spinal cord
- A44.9 Unspecified partial extirpation of spinal cord
- A45.1 Stereotactic chordotomy of spinal cord
- A45.2 Open chordotomy of spinal cord NEC
- A45.3 Myelotomy of spinal cord
- A45.4 Open biopsy of lesion of spinal cord
- A45.5 Removal of foreign body from spinal cord
- A45.6 Open aspiration of lesion of spinal cord
- A45.8 Other specified other open operations on spinal cord
- A47.1 Needle destruction of substantia gelatinosa of cervical spinal cord
- A47.2 Radiofrequency controlled thermal destruction of spinothalamic tract
- A47.3 Percutaneous chordotomy of spinal cord
- A47.8 Other specified other destruction of spinal cord
- A48.1 Biopsy of lesion of spinal cord NEC
- A48.2 Aspiration of lesion of spinal cord
- A48.3 Insertion of neurostimulator adjacent to spinal cord
- A48.4 Attention to neurostimulator adjacent to spinal cord NEC
- A48.6 Removal of neurostimulator adjacent to spinal cord
- A48.7 Insertion of neurostimulator electrodes into the spinal cord
- A48.8 Other specified other operations on spinal cord
- A49.1 Freeing of spinal tether NEC

- A49.2 Closure of spinal myelomeningocele
- A49.3 Closure of spinal meningocele
- A49.4 Complex freeing of spinal tether
- A49.8 Other specified repair of spina bifida
- A49.9 Unspecified repair of spina bifida
- A51.1 Extirpation of lesion of meninges of spinal cord
- A51.2 Freeing of adhesions of meninges of spinal cord
- A51.3 Biopsy of lesion of meninges of spinal cord
- A51.8 Other specified other operations on meninges of spinal cord
- A51.9 Unspecified other operations on meninges of spinal cord
- A53.1 Cerebrospinal syringostomy
- A53.3 Creation of syringoperitoneal shunt
- A57.1 Extirpation of lesion of spinal nerve root
- A57.6 Reimplantation of spinal nerves into spinal cord

Posterior eye surgical procedures that are regarded as high-risk posterior segment eye surgery

Orbit (C01 to C08)

- C01 Excision of eye
- C03 Insertion of prosthesis of eye
- C04 Attention to prosthesis of eye

These orbital operations are only included if the surgery or implant is likely to come into contact with the optic nerve or retinal tissue (for example, evisceration of the eye and intra-orbital implant).

Operations on optic nerve (A29.1 to A36.4)

- A29.1 Excision of lesion of optic nerve
- A30.1 Repair of optic nerve
- A32.1 Decompression of optic nerve
- A34.1 Exploration of optic nerve
- A36.4 Radial Optic Neurotomy

Sclera and iris (C52 to C65)

C54 Buckling operations for attachment of retina

Retina, other parts of eye and anaesthetics (C79 to C90)

- C79 Operations on vitreous body, only when this involves potential contact with the posterior hyaloid face. For example:
  - Code C7910 for vitrectomy via anterior approach and code C7923 for intravitreal injections are specifically excluded because they are unlikely to come into contact with the posterior hyaloid face.
  - Code C7920 and code C7922, which potentially could come into contact with the hyaloid face, are included.
- C80 Operations on retinal membrane
- C81 Photocoagulation of retina for detachment, only when the retina is handled directly
- C82 Destruction of lesion of retina, only when the retina is handled directly
  - Code C82.4 for insertion of radiotherapy plaques is specifically excluded.
- C83 Translocation of retina
- C84 Other operations on retina

- C85 Fixation of retina
- C86 Other operations on eye
- C88 Destruction of subretinal lesion
- C89 Operations on posterior segment of eye

Update information

Minor changes since publication

January 2026: Interventional procedures guidance 666 has been migrated to HealthTech guidance 535. The recommendations and accompanying content remain unchanged.

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Endorsing organisation

This guidance has been endorsed by Healthcare Improvement Scotland.