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MANAGING CJD/VCJD RISK IN NEUROSURGERY

INTRODUCTION AND CORE MESSAGE

Over the last 50 years, as understanding of Creutzfeldt-Jakob Disease (CJD) has developed, the expectations on neurosurgeons to protect patients against iatrogenic disease have grown. Pragmatic guidance has been produced based on assumptions that the science is correct. Precautions are expected in order to reduce an uncertain and far distant future risk. The consequence of ignoring the issue, or getting it wrong, could be a widespread self-sustaining epidemic which would be a human disaster.

Human prion diseases are not contagious, but can be transmitted from one individual to another by the inoculation with or consumption of infected tissues (including blood). The highest levels of infectivity are found in the central nervous system (CNS) during the clinical phase of the illness, and there is experimental evidence to indicate that CNS tissues may be able to transmit disease for several years before the symptoms of CJD finally become apparent.

Infectivity is closely related to the presence of abnormal prion protein in tissues. Reusable neurosurgical instruments may carry residues of baked-on protein which, if contaminated by abnormal prion protein from affected CNS tissues, is not fully removed by current decontamination methods. This residuum may be sufficient to infect subsequent patients on whom these neurosurgical instruments are used.

It is vitally important to do everything possible to protect the well-being of those not otherwise at risk of CJD/vCJD by avoiding their exposure to potentially contaminated intradural instruments.

THE DEPARTMENT OF HEALTH GUIDANCE IS LARGELY ABOUT CLINICAL CJD AND KNOWN HISTORIC EXPOSURES TO INFECTION

Patients with CJD or where a primary diagnosis of CJD cannot be excluded before intradural surgery (including pituitary surgery) pose a risk to others. They should be identified prior to surgery and should have precautions taken with the surgical instruments used, to prevent the infection of other patients.

These precautions also apply to patients with an increased risk of developing CJD through their past healthcare, because of a genetic risk of inherited prion disease, and those where the risk cannot be determined prior to the procedure.

The numerical risk is small, but real and taken very seriously. If any of these patients slip through pre-operative screening and are later discovered, subsequent patients who have been exposed to the potentially contaminated surgical instruments are traced and informed they are at risk and must take public health precautions to reduce the likelihood of infecting others.

THE NICE GUIDANCE 2006 IS PRIMARILY CONCERNED WITH VARIANT CJD (vCJD) AND THE GENERAL POPULATION

People living in the UK before 1997 are considered to be at greater risk of developing vCJD in later life due to dietary exposure to Bovine Spongiform Encephalopathy (BSE) than those born after this date. There is evidence of abnormal prion protein in gut-related lymphoid tissue in roughly 1 in 2,000 (0.05%) of the older population, with no way currently of identifying those affected or knowing the long-term implications for them. This underpins the need to ensure neurosurgical instruments remain within their individual sets. It is the only practical means to limit the surgery-associated spread of vCJD within the population born before 1997. Provided all instruments in a contaminated set remain together, only one subsequent patient at a time is exposed. If contaminated Instruments migrate to other sets the number of patients exposed to them is greatly increased, amplifying the consequences.

In order to limit the risk of an iatrogenic self-sustaining second epidemic, and eventually to abolish the risk of vCJD, pre-97 instruments must not be used for intradural neurosurgery (including pituitaries) in

those born after 1996. Separate "post-96" instruments for use only in this younger age group "born after 1996" should be rigorously guarded.

A review of the NICE guidance is scheduled – until the outcome of this review is known, the advice set in the 2006 guidance remains current and is reflected throughout this document. .

THE PURPOSE OF THIS GUIDANCE

N1. This Guidance was developed jointly by the Department of Health (DH) Advisory Committee on Dangerous Pathogens Transmissible Spongiform Encephalopathy Subgroup (ACDP TSE SG), the National Institute for Health and Care Excellence (NICE), and the Society of British Neurological Surgeons (SBNS).

N2. It links advice from the DH Guidance on "Minimising the transmission risk of CJD and vCJD in healthcare settings"[1](https://www.gov.uk/government/publications/guidance-from-the-acdp-tse-risk-management-subgroup-formerly-tse-working-group), with the NICE Guidance on "Patient safety and reduction of risk of transmission of CJD via interventional procedures"[2] (http://www.nice.org.uk/IPG196) in a single document. It describes the current status of CJD in the UK and provides practical advice for neurosurgeons and neurosurgical units to implement these guidelines for both elective and emergency cases.

THE RELEVANCE OF CJD AND vCJD TO NEUROSURGICAL PRACTICE

N3. Worldwide, several hundred people have died from CJD acquired through healthcare, including the use of products manufactured from pooled human growth hormone sourced from pituitary glands (>200 cases) or cadaveric dura mater (>200 cases), contaminated by as little as one affected donor. Deaths are still occurring 30 years after these products were banned. Transmission from re-used EEG needles or neurosurgical instruments has caused up to 7 cases[3,4]. To date there has been no known iatrogenic transmission of variant CJD (vCJD) following surgery, but transmission via blood transfusion (4 instances) and UK plasma used to produce Factor VIII (1 instance) has occurred. Intradural neurosurgery (including pituitaries) has the highest surgical risk of transmitting iatrogenic CJD.

N4. All neurosurgical units occasionally operate on patients with an unknown neurological diagnosis that might turn out to be CJD, with previous neurosurgical or medical treatments that might increase their risk of CJD, with a family history or genetic predisposition to CJD, or with asymptomatic carriage of CJD. The risk of onward transmission posed by these patients may not be apparent at the time.

N5. The risk of causing iatrogenic infection with CJD is low in comparison with other more immediate neurosurgical risks. However, the risk is real and cannot be ignored. Currently there is no reliable test to identify infected but asymptomatic persons, and no effective technology for prion removal from surgical instruments. The main strategy to prevent person to person spread through surgery remains avoidance of exposure to affected instruments by the risk reduction approach outlined in this document.

N6. The emergence of vCJD in the 1990s following an epidemic of BSE in cattle in the 1980s caused widespread concern. Measures to prevent BSE entering the food chain culminated in a reinforcement of the ruminant feed-ban in 1996 and are credited with avoiding a much larger epidemic of vCJD. On a precautionary basis, it is assumed that anybody born in the UK before 1997 has potentially been exposed to vCJD through their diet but that this exposure had largely ceased by 1996. There are a number of public health measures in place to protect the growing post-96 cohort of the population from potential exposure through healthcare or blood products[5], including guidance for neurosurgical procedures[1,2].

N7. The retrospective discovery of a patient with the possibility to infect others with CJD is a serious event. The incident must be reported, the instruments tracked and destroyed, and patients operated on

subsequently using any of these instruments traced and informed that they are regarded as at risk of CJD for public health purposes. They must declare this in future and follow other public health restrictions. The upset and adverse attention this causes is distressing and disruptive for the patients, the surgeon, and the unit involved, so should be avoided if at all possible by careful risk assessment.

N8. Consequences also follow for patients born after 1996 who have intradural operations performed with instruments previously used in those born before 1997. They should be informed of their exposure to the general population risks of vCJD. Thereafter any future intradural operations should be performed with single-use instruments or with new re-usable instruments or post-96 instruments that are subsequently quarantined and kept for use exclusively on that patient or added to the pre-97 stock.

BACKGROUND

THE EPIDEMIOLOGY OF CJD IN THE UK

N9. CJD is one of a group of invariably fatal neurodegenerative diseases caused by pathological accumulation in the brain of an aberrant form (PrP^{Sc}) of the normal cell glycoprotein, prion protein (PrP).

N10. Sporadic CJD is the most common form. It occurs worldwide with an incidence of 1-2 deaths per million population per year. It mostly affects people over the age of 60. The clinical disease is characterised by rapidly progressive dementia, myoclonus, and ataxia followed by mute immobility and death. About 100 people in the UK die from sporadic CJD each year.[6] In addition there are approximately 5-10 deaths from inherited prion disease and 1-2 from iatrogenic CJD (linked to historic treatment with pooled products such as human growth hormone or contaminated dura).

N11. Variant CJD typically affects young adults in their late 20s. The clinical course lasts more than a year and is dominated by psychiatric and sensory disturbances in the early stages. There have been 178 deaths from vCJD in the UK. New diagnoses have declined since 2000 and are currently occurring at a very low rate, with 0-1 new diagnoses each year since 2010[6].

N12. The incubation periods associated with acquired forms of CJD are variable, but can be decades, especially for low doses introduced at peripheral sites.[3] Around 5,000 people have been identified and informed that they are at increased risk of developing CJD because of their previous medical treatment, such as treatment with certain UK sourced plasma products or human-derived growth hormone. They have been asked to take public health precautions, including providing this information at pre-surgical assessment so that the management of neurosurgical instruments can be arranged.

N13. Of the general population exposed to BSE through their diet, the number who may be infected with vCJD but yet to show symptoms is unknown. Fears of a large-scale epidemic have lessened, but the true prevalence of asymptomatic vCJD (infected and infectious but not yet clinically apparent) is uncertain. Population-based studies looking for abnormal prions in resected appendix tissue indicate that 1:2,000 of the population born before 1997 may be affected.[7]

N14. The need to assess patients for the risk of transmitting CJD and destroy instruments when there is any doubt may seem disproportionate. However, the likelihood of iatrogenic infection causing a self-sustaining second epidemic is judged sufficient to justify the difficulties involved in assessing risk and withdrawing instruments from use.

THE TISSUE DISTRIBUTION AND INFECTIVITY OF CJD/vCJD

N15. Observations of naturally occurring disease and primary experimental infection clearly identify central nervous tissue as carrying the highest level of abnormal prion proteins and as the most infectious source of CJD.

N16. For sporadic CJD, infectivity is largely confined to central nervous tissue, and in sufficient concentrations to cause iatrogenic transmission for several years before clinical symptoms. The same infectivity is assumed for other forms of CJD in their pre-clinical stages.

N17. For vCJD, infectivity is also detected within peripheral lymphoreticular tissues and blood, though the concentration of prion proteins in peripheral tissues is considerably less than within central nervous tissue. For vCJD it is assumed that infectivity from peripheral tissues is present in low to medium titre throughout the incubation period, and from central nervous tissues in high titre during the latter part of the pre-clinical phase of disease.

N18. Operations involving the central nervous system including cranial nerves, pituitary, and posterior eye carry the highest risk of transmitting CJD/vCJD.

N19. Decontamination of re-usable instruments is a cornerstone of safe surgery. Despite this, most surgical instruments carry a film of baked-on protein residue, which in the case of CJD is resistant to conventional decontamination methods and remains demonstrably infectious.[8] More effective decontamination methods are being developed but at present even the best existing practice does not completely remove prion protein from instruments. This guidance recognizes these shortcomings and relies principally on the avoidance of patient contamination rather than instrument decontamination.

N20. Modelling suggests that surgical transmission could potentially act as a significant multiplier of the initial primary outbreak of dietary associated vCJD.[9] This assessment and the lack of a reliable method to identify sporadic, iatrogenic, or vCJD infection during the asymptomatic period continue to justify the difficulties of assessing the risk for individual patients and withdrawing potentially contaminated surgical instruments from use and keeping neurosurgical sets together to limit the possibility for multiple iatrogenic exposure.

DEFINITIONS

POPULATION GROUPS AT DIFFERENT RISKS OF DEVELOPING OR TRANSMITTING CJD/vCJD

There are four population groups to consider in order to limit the risks of accidental transmission of CJD by neurosurgery.

GROUP ONE - SYMPTOMATIC PATIENTS WITH CJD, OR FOR WHOM A DIAGNOSIS OF CJD IS BEING CONSIDERED OR CANNOT BE EXCLUDED

GROUP TWO - ASYMPTOMATIC PATIENTS WITH AN INCREASED RISK OF DEVELOPING CJD

N21. Risk management for both these groups in healthcare and community settings is described in detail within the DH guidance.[1] The guidance on management of neurosurgical instruments is similar for all those with a known diagnosis or an increased risk of developing CJD. It is useful however, to make a distinction between:

N22. Symptomatic patients

- Definite, probable or possible CJD: Patients who fulfil the diagnostic criteria for definite, probable or possible CJD or vCJD rarely undergo neurosurgery except occasionally for histological diagnosis. The clinical disease is characteristic and short-lived, and neurosurgery for reasons other than diagnosis is rarely indicated.
- Undiagnosed neurological disease: Occasionally patients with an undiagnosed neurological disease, where CJD has not been excluded, undergo neurosurgical treatment including diagnostic biopsies. Precautions for the instruments involved and recommendations for storage of reference tissues should be followed.[1 – Annex I]

N23. Asymptomatic patients "at increased risk"

These are people who have no clinical symptoms of CJD but who are "at increased risk" of developing the disease because of their family or medical history.

- Patients "at increased risk" from inherited prion disease (genetic CJD)
  Nearly all people with a disease-specific mutation and 50% of those who have a blood relative with this mutation but who have not themselves been tested will develop inherited prion disease. They are likely to be infectious long before they become clinically unwell.
- Patients "at increased risk" because of their medical history
  People may develop iatrogenic CJD after exposure to infection through:
  - receiving contaminated blood, blood components or plasma products;
  - treatment with contaminated dura mater grafts;
  - treatment with human-derived hormones derived from pituitary glands such as human growth hormone or gonadotrophin;
  - surgery or other invasive medical procedures using contaminated instruments.

  Many asymptomatic patients "at increased risk" (and their GP or a medical specialist in charge of their long term care) are already informed of their status. These individuals are asked to follow public health measures in order to prevent potentially infecting others. For patients who may not have been informed prospectively, for example dura mater graft recipients, the pre-surgical risk assessment process and screening questions should identify the risk.[1 - Annex J]

GROUP THREE - THOSE BORN BEFORE 1997

N24. The entire population born in the UK before 1997 is considered to have been exposed through diet to some risk of developing vCJD after an unknown latent period that may extend to several decades. The current evidence suggests that around 1:2,000 of the population born before 1997 have identifiable levels of abnormal prion protein in their peripheral lymphoid tissues and may be incubating the disease. There is currently no means to identify these individuals prospectively.

N25. The 2012 ACDP position statement on occurrence of vCJD and prevalence of infection in the UK population makes precautionary assumptions; that further clinical cases of vCJD may appear after much longer clinical incubation periods than those seen so far; that presence of abnormal prions is indicative of vCJD infectivity and that, while asymptomatic, these individuals represent possible sources of further onward infection.[10]

N26. In 2004 the Department of Health commissioned the National Institute for Health and Care Excellence(NICE) to consider ways of reducing the risks of a potentially self-sustaining epidemic occurring due to contamination of surgical instruments following procedures being carried out on people infected with, but yet to develop clinical symptoms of, vCJD. The resulting NICE guidance published in 2006 identified a number of steps that together, while not preventing all infections, should limit iatrogenic exposure and the risks of perpetuating this disease.

N27. NICE assumed that its guidance might be short-term until effective decontamination and sterilisation techniques became available for CJD. This cannot yet be assured and the NICE guidance remains extant.

GROUP FOUR - THOSE BORN AFTER 1996 AND AT REDUCED RISK OF vCJD.

N28. People born after 1996 are less likely to have been exposed to BSE through food. They are considered to have a reduced risk of developing vCJD. This group are also free from risk of CJD from human dural grafts or human hormone treatment. The NICE guidance aims to protect this population against vCJD infection by not sharing neurosurgical instruments with the older population.

Table 1 describes the classification of the risk status of symptomatic patients and asymptomatic patients "at increased risk" according to the DH guidance.[1 – Part4]

Table 1: Patient groups	
Symptomatic patients	• Patients who fulfill the diagnostic criteria for definite, probable or possible CJD or vCJD (see [1 - Annex B] for diagnostic criteria) • Patients with neurological disease of unknown aetiology, who do not fit the criteria for possible CJD or vCJD, but where the diagnosis of CJD cannot be excluded.
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 *(informed).* • Individuals who have a blood relative known to have a genetic mutation indicative of genetic CJD *(informed).* • Individuals who have or have had <u>two or more</u> blood relatives affected by CJD or other prion disease *(these individuals may not be aware of their CJD risk).*
Patients "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 *(informed).*
Patients "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 *(most individuals treated in the UK have been informed of their CJD risk).* • 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) *(these individuals may not be aware of their CJD risk).* • 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 *(informed).* • Individuals who have received an organ or tissue from a donor infected with CJD/vCJD or "at increased risk" of CJD/vCJD *(informed).* • Individuals who are known at referral to have received blood or blood components from 300 or more donors since January 1990. The figure of 300 is a guide, not a precise cutoff. Only a small proportion of transfusion recipients have received this quantity, predominantly for the treatment of haematological diseases *(these individuals may not be aware of their CJD risk).* • Individuals who have given blood to someone who went on to develop vCJD; and other recipients of blood from those donors *(informed).* • Individuals who have been treated with certain implicated UK sourced plasma products between 1990 and 2001, mainly to treat inherited bleeding disorders *(informed).*

Informed: these patients and their GP, or specialist clinician involved in their ongoing care, such as a haematologist, have been informed about their increased risk of CJD.

Box 1: NICE GUIDANCE

In summary NICE Guidance (2006) recommends

i) **Ensuring that instruments that come into contact with high-risk tissues do not migrate between sets and that supplementary instruments are kept faithful to a single parent set.**
Rationale: When contaminated instruments are kept together in a set they can only infect individuals who follow sequentially. Instruments migrating to other sets expose a greater number of individuals.

ii) **For neuroendoscopy: rigid neuroendoscopes should be used wherever possible and single-use accessories should be used in all cases.**
Rationale: Decontamination of flexible endoscopes is less effective than rigid endoscopes. The evidence on cost effectiveness related to the risk of possible CJD transmission supports the use of single-use neuroendoscopy accessories.

iii) **Provision and use of a separate pool of new neuroendoscopes and reusable surgical instruments for high-risk procedures for individuals born after 1996 (who are unlikely to have been exposed to BSE in the food chain or vCJD through a blood transfusion). These instruments should not be used for patients born before 1997**
Rationale: It is not known how many of those born before 1997, and exposed to the risk of developing vCJD through their diet may eventually develop clinical disease. It is possible that many of those infected may remain asymptomatic for several decades. Since we cannot yet identify individuals with sub-clinical disease the risk of cross-infection between the older and younger age groups is limited by keeping their neurosurgical instruments separate.

NICE Guidance 2006 Quick Reference Guide and Full Guidance[2]

INSTRUMENT GROUPINGS

RESERVED POST-1996 INSTRUMENT STOCK

N29. The NICE guidance emphasises the need for a separate pool of neuroendoscopes and reusable surgical instruments for intradural procedures on patients born after 1996.

N30. Post-96 instruments / neuroendoscopes should be purchased specifically for use in the younger age group, born after 1996 and kept solely for them. It is safe to reuse instruments within this population provided there are no additional risk factors, as described in Table 1 .

N31. There will be a small number of patients born after 1996 who were operated on using pre-1997 instruments before the NICE guidance was issued in 2006. For these patients, further intradural procedures should

- Use single-use instruments or
- Use new reusable or post-1996 instruments and either retain them for sole use on this patient, or afterwards add to the pre-1997 stock.

N32. If instruments from the reserved post-1996 stock are used deliberately or by mistake in a patient born before 1997, they should not be returned to the post-96 stock, but may continue to be used as part of the pre-97 stock.

PRE-1997 INSTRUMENT STOCK

N33. Instruments used on patients born before 1997 should not be used in patients born after 1996. If this happens, the patient, or their parents/guardians, must be informed and should declare this before further intradural procedures so that efforts may be made to reduce the likelihood of contaminating the post-96 instruments (see N31).

N34. The individual risk to these patients is very low, and this information does not mean they are "at increased risk" of developing CJD as described in Table 1.

SINGLE-USE INSTRUMENTS

N35. NICE considered single-use instruments to be the safest option with regard to preventing transmission but was concerned about manufacturing quality assurance and cost-effectiveness. Much industry effort has been made to deliver high-quality reasonably-priced single-use instruments.

N36. Single-use instruments/neuroendoscopes are recommended for use where they are available to a suitable quality and patient care is not compromised as a result.

PRACTICAL MANAGEMENT OF NEUROSURGICAL PATIENTS

N37. It is important to identify people with a known or suspected diagnosis of CJD or an increased risk of developing CJD, to ensure that instruments used in these cases are not re-used on others.

PATIENTS WITH PROGRESSIVE NEUROLOGICAL DISORDERS

N38. Patients undergoing diagnostic biopsy of central nervous tissue where CJD cannot be reliably excluded prior to histological confirmation should be considered to be at risk of transmission and precautions taken, irrespective of their answers to the risk assessment questions.

N39. A protocol for the management of instruments and tissues from brain biopsy on patients with progressive neurological disorders has been produced by the Department of Health.[1 - Annex I]

PRE SURGICAL RISK ASSESSMENT

N40. The purpose of the pre surgical risk assessment is to determine which of the four population groups an individual falls into.

N41. All patients undergoing intradural surgery (including pituitaries) should be regarded as potentially infective for CJD unless additional risks can be excluded. If additional risks can be excluded, the patient's date of birth determines which instruments to use.

N42. Risk assessment should be undertaken using the CJD risk assessment questions, with supplementary clarification to define these risks where there is doubt.[1 - Annex J]

N43. Patients may be able to provide the information required directly or through their family or General Practice. In the event that the information is not available the senior operating surgeon must make an assessment of the risk factors based on the clinical presentation.

N44. **CJD RISK ASSESSMENT QUESTIONS for patients undergoing intradural surgery (including pituitaries)[1 - Annex J]**

All patients should be asked:

- **Have you ever been notified that you are at risk of CJD or vCJD for public health purposes?**
  Around 5,000 individuals have been informed that they are at risk of CJD or vCJD, and are expected to provide this information before a surgical procedure. Their GP or the specialist clinician involved in their ongoing care, such as a haematologist, has also been informed (see Table 1).

Further questions which aim to identify patients who are not aware of their risk (see Table 1):

- **Have you a history of CJD or other prion disease in your family?**
  Further questions should ascertain whether the patient has 1) had a genetic test that indicates they have a risk of developing prion disease, 2) a blood relative with a genetic mutation indicative of genetic CJD/prion disease or 3) two or more relatives affected by CJD/other prion disease. If the answer to all three of these questions is no or not known, then no further investigation is required.

This should differentiate these from the larger number of patients where a single relative has died from a non-genetic form of CJD, so are not themselves at added genetic risk.

- **Have you ever received growth hormone or gonadotropin treatment?**
  Most patients treated with human-derived hormones in the UK have been informed of their CJD risk. Some may have been treated when they were children and some may have received these products abroad. Further questions should ascertain whether the hormone was derived from human pituitary glands, the year of treatment and whether treatment was received in another country.
- **Have you ever had surgery on your brain or spinal cord?**
  Patients with previous brain or spinal surgery should be considered at increased risk if, in the opinion of the patient's consultant, a dural patch may have been used and the use of human-derived dura mater cannot be ruled out by reference to the medical records. For example: spinal surgery involving an expansion duraplasty; foramen magnum decompression; cranial surgery involving extensive dural resection and repair (eg meningioma resection); major trauma craniotomy; basal dural repair. The risk is from the use of human-derived dura mater before 1992. Of the over 200 cases seen to date, almost all have been associated with Lyodura. The mean incubation period is 12 years but has been as long as 30 years, and new cases still occur.[3]

N45. A yes answer to any of these questions, or an inability to answer any of these questions, requires that the patient be treated provisionally as a high risk for CJD transmission while further clarification is sought. **Surgery should not be delayed but should be undertaken with precautions to avoid iatrogenic spread unless any subsequent clarification proves that the patient does not pose a risk and precautions are therefore unnecessary.**

N46. If a patient, who is not already aware of their increased risk, is identified at the presurgical assessment they should be referred to their GP, who will need to inform them of their increased risk of CJD or vCJD and provide them with further information and advice. This is available from the CJD section at Public Health England, https://www.gov.uk/government/publications/cjd-information-leaflets-for-patients-and-healthcare-professionals. Tel: 020 327 6090. Patients who are at increased risk of genetic forms of CJD should be offered the opportunity of referral to the National Prion Clinic, based at the National Hospital for Neurology and Neurosurgery, Queen Square, London: http://www.nationalprionclinic.org/. Patients who are at increased risk of sporadic CJD due to receipt of human-derived growth hormone should be offered the opportunity of referral to the UCL Institute of Child Health, London

EMERGENCY ADMISSIONS

N47. Surgery should not be postponed to permit the risk assessment questions to be answered. In an emergency, or when it is not otherwise possible to assess risk, the patient should be treated as potentially infective for CJD and either single-use instruments should be used, or the instruments used should be quarantined for use on this patient alone, then returned to stock or destroyed as appropriate once the risk has been established.

CJD RISK MANAGEMENT – GENERAL PRINCIPLES

Dura Mater – the boundary between high and low risk procedures

N48. Although the past use of contaminated, pooled cadaveric dura is associated with iatrogenic CJD, dura mater itself is not a high infectivity tissue, even in a patient with a clinical diagnosis of CJD.

N49. Consequently only those instruments which have been used inside the dura and therefore coming in direct contact with high risk tissues (brain and spinal cord) need be identified and kept separate from other instruments. Such instruments should be kept separate from other instruments on the set to reduce the risk of cross-contamination and either incinerated after use or quarantined for re-use exclusively on the same patient.

Instrument management

N50. All instruments should be tracked and kept in their individual set. Supplementary instruments should be single-use or allocated to a single parent set.

Single-use instruments

N51. Where available, satisfactory single-use instruments of similar quality to reusable instruments represent the simplest safeguard against the risk of iatrogenic infection. When single-use surgical instruments are used they should be kept separate from re-usable instruments and disposed of at the end of the procedure

Decontamination

N52. Current methods for decontamination of re-usable instruments are not able to fully remove all protein residue. New technologies are being developed, and it is hoped that these will enable more efficient decontamination in future.

N53. Preventing instruments from drying out can substantially reduce prion protein adherence and infectivity. Instruments should be kept moist after use and cleaned as soon as possible.

N54. There is no need for dedicated decontamination facilities for instruments reserved for use in the younger age group born after 1996. It is good practice to process these instruments separate from pre-97 instruments but there is no absolute need to do so as there is no evidence of cross-contamination in washers.

N55. Further information can be found in the Department of Health's guidance on decontamination of surgical instruments (HTM 01-01)[11] and in ACDP's General principles of decontamination and waste disposal[1 – Annex C].

CJD RISK MANAGEMENT – SPECIFIC EXAMPLES

GROUP ONE -- Patients with CJD, with an uncertain diagnosis or an unknown risk for whom a diagnosis of CJD is being considered or cannot be excluded

N56. For patients where CJD is known or suspected, and for patients undergoing intradural surgery (including diagnostic biopsies) where a primary diagnosis of CJD cannot be excluded, **including patients unable to complete the risk assessment in an emergency or when it is not otherwise possible to assess risk, the patient should be treated as a potential risk of transmission**. The instruments should be single-use, or quarantined (for use in that patient alone) until risk status is confirmed. If CJD risk is positively excluded the instruments may be returned to stock; if the risk cannot be excluded they should be kept quarantined (for use in that patient alone) or destroyed.

GROUP TWO -- Patients at increased risk of developing CJD

N57. Patients at increased risk of developing CJD (table 1), **including where the risk is suspected but not certain**. The instruments should be single-use, or quarantined (for use in that patient alone) until risk status is confirmed. If CJD risk is positively excluded the instruments may be returned to stock; if risk cannot be excluded they should be kept quarantined (for use in that patient alone) or destroyed.

GROUP THREE -- Patients born before 1997

N58. Patients born before 1997, not otherwise at risk of CJD, should be operated on using re-usable instrument sets and neuroendoscopes reserved for this age group.

GROUP FOUR -- Patients born after 1996 at reduced risk of vCJD

N59. Patients born after 1996 with no history of previous intradural surgery undertaken with pre-1997 instruments or other identifiable risk factors for CJD (table 1) should be operated on using the separate stock of re-usable instruments and neuroendoscopes reserved for this age group.

Table 2: The use and management of neurosurgical instruments by patient group. Irreversible actions such as permanent disposal of instruments should not be done until the diagnostic status is clear.

Patient group	Use…	Instrument management
Patients with or at increased risk of developing CJD (Groups 1&2)	Single-use	Destroy
	Standard pre 1997 stock	Quarantine or keep for exclusive use on this patient OR Destroy *For patients with an unclear or unknown neurological diagnosis or increased risk of developing CJD – permanent disposal of quarantined instruments should wait until the diagnosis is certain or risk status is confirmed. If CJD diagnosis or risk is later positively excluded they may be returned to stock*
Those born before 1997 (Group 3)	Standard pre 1997 stock	Decontaminate and return to standard stock
Those born after 1996 (Group 4)	Reserved post 1996 stock	Decontaminate and return to reserved stock

SUMMARY

1. Patients undergoing extradural surgery pose only a small risk of iatrogenic transmission (even in the case of an inadvertent CSF leak uncontaminated by brain or spinal cord), and instruments may be re-used.
2. All patients undergoing intradural surgery (including pituitary surgery) should be assessed for the risk of CJD.
3. If risk is demonstrated pre-operatively then instruments used for intradural surgery should be single-use, or quarantined for use in that patient alone, or destroyed.
4. If risk cannot be established pre-operatively then instruments used for intradural surgery should be single-use, quarantined for use in that patient alone until risk is excluded, or destroyed.
5. If risk can be excluded pre-operatively or post-operatively then instruments used for intradural surgery may be returned to stock and re-used.
6. For patients born before 1997 and not otherwise at risk of CJD, reusable instruments should be from the stock of “pre-97” instruments.
7. For patients born after 1996 and not otherwise at risk of CJD, reusable instruments should be from a reserved stock of “post-96” instruments kept only for that age group. Where this has not happened the patient or their parent/guardian should be informed.
8. For patients born after 1996 who have undergone past intradural surgery with older “pre-97” instruments. The simplest solution is to use single-use instruments where available and of suitable quality. Alternatively new re-usable instruments, or post-96 instruments. may be used and quarantined for use in that patient alone or added to the general “pre-97” stock.
9. If reserved post-96 instruments are used deliberately or inadvertently in a patient born before 1997, they should be moved to the pre-97 stock.
10. The later discovery that a neurosurgical patient has developed CJD or has a recognised risk of developing CJD, and the instruments have been re-used, is a CJD incident. The instruments used should be tracked and quarantined. Theincident should be reported to the local infection prevention and control department for contact tracing and notification etc.

These guidelines have been produced as an aid to good clinical practice. They present risk management advice, based on published evidence and expert opinion. The ultimate judgment regarding management of a particular situation must be made by the senior operating surgeon based on the best information that is available to them. It is acknowledged that decisions must sometime be taken, where information is poor or lacking and that problems have been reported tracking individual instruments through the sterile services system, especially when this is located offsite.

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   Annex A1: Distribution of transmissible spongiform encephalopathy infectivity in human tissues and body fluids
   Annex B: Diagnostic criteria
   Annex C: General principles of decontamination and waste disposal
   Annex I: Outline protocol for management of instruments and tissues from brain biopsy procedures on patients with progressive neurological disorders
   Annex J: Assessment to be carried out before surgery and or endoscopy to identify patients with, or at risk of, CJD or vCJD
   Part 4 Infection control of CJD, vCJD and other human prion diseases in healthcare and community settings

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Appendix 1: Patient management algorithm