

ANNEX J

Assessment to be carried out before surgery and/or endoscopy to identify patients with, or at increased risk of, CJD or vCJD

Summary of advice (revised January 2014)

Annex J provides a clear and pragmatic way of assessing CJD and vCJD risk prior to surgery or endoscopy. Certain groups of patients have been informed that they are at increased risk of CJD or vCJD. Therefore it is recommended that all patients about to undergo any surgery or endoscopy should be asked if they have ever been notified as at increased risk of CJD or vCJD.

This recommendation is outlined in [paragraphs J1 and J2](#).

In addition, patients undergoing surgery or neuro-endoscopy which may involve contact with tissues of potentially high level TSE infectivity (“high risk tissues”) should, through a set of detailed questions, be assessed for their possible CJD/vCJD risk exposure. These questions are outlined in [Table J1](#) and [paragraphs J3 to J6](#).

Previous revision date: January 2013

Changes new to this edition:

Date	Change	Notes
January 2014	Alignment of the list of people considered at increased risk of vCJD with that contained in Part 4.	This change affects paragraph J14.
January 2014	Change of terminology from “infection control” to “infection prevention and control”	Changed throughout the document as appropriate

Recommendation for all surgical and endoscopy patients

- J1. The CJD Incidents Panel has identified a number of individuals or groups who are at increased risk of CJD or vCJD ([see paragraphs J14 – J18](#)).

At a local level arrangements should be put in place to ensure that patients who have been notified they are at increased risk of CJD/vCJD are identified before surgery or endoscopy, to allow appropriate infection prevention and control procedures to be followed.

All patients about to undergo **any** elective or emergency surgical or endoscopic procedure should be asked the question:

“Have you ever been notified that you are at increased risk of CJD or vCJD for public health purposes?”

- J2. The actions to take following the patient’s response to the above question are:

Patient’s response	Action
No	Surgery or endoscopy should proceed using normal infection prevention and control procedures unless the procedure is likely to lead to contact with high risk tissue.
Yes	<p>Please ask the patient to explain further the reason they were notified.</p> <p>Special infection prevention and control precautions should be taken for all surgery or endoscopy involving contact with medium or high infectivity tissues (see Annex A1) and the local infection prevention and control team should be consulted for advice.</p> <p>Part 4 of this Guidance provides advice on the precautions to be taken during the treatment of patients with or at increased risk of CJD or vCJD, and Annex F provides information on endoscopic procedures.</p> <p>The patient’s response should be recorded in their medical notes for future reference.</p>
Unable to respond	Surgery or endoscopy should proceed using normal infection prevention and control procedures unless the procedure is likely to lead to contact with high risk tissue. If this is the case, please refer to the additional recommendations for high risk procedures from paragraph J3 onwards, with particular reference to paragraphs J7 – J10 .

Additional recommendations for surgery and neuro-endoscopy which may involve contact with high risk tissue only

N.B. These additional recommendations are only applicable to those assessing patients in neurosurgical and ophthalmic surgical departments for intradural and posterior ophthalmic surgical procedures. With regards to endoscopy, these additional recommendations are only applicable to those assessing patients for intradural neuro-endoscopic procedures.

Procedures should not be delayed whilst information is being collected, and clinicians should be careful not to prejudice overall patient care.

J3. As well as asking all patients whether they have been notified as being at increased risk of CJD/vCJD, clinicians assessing patients for procedures that involve contact with high risk tissues should ask supplementary questions ([as outlined in Table J1](#)) to assess further their CJD/vCJD risk. If a patient has answered 'yes' to the question in paragraph J1 there is no additional need to ask the questions in Table J1 – the patient's risk status has been established.

J4. Tissues assumed or proven to have high level infectivity for CJD or vCJD are:

- Brain
- Spinal cord
- Implanted dura mater grafts prior to 1992
- Cranial nerves, specifically:
 - the entire optic nerve
 - only the intracranial components of the other cranial nerves
- Cranial nerve ganglia
- Posterior eye, specifically:
 - posterior hyaloid face
 - retina
 - retinal pigment epithelium
 - choroid
 - subretinal fluid
 - optic nerve
- Pituitary gland

[Annex A1](#) gives further advice on CJD/vCJD tissue infectivity

J5. [Table J1](#) outlines recommended questions to assess CJD/vCJD risk. It is recommended that patients are asked these questions prior to elective or emergency surgical or neuro-endoscopic procedures likely to involve contact with tissues of potentially high infectivity. [Paragraph J6](#) outlines the steps to take based on the patient's responses. [Appendix B](#) is an information sheet for pre-surgical patients undergoing surgery or neuro-endoscopy on high risk tissues about the questions they will be asked.

Table J1 – CJD risk questions for patients about to undergo elective or emergency surgical or neuro-endoscopic procedures likely to involve contact with tissues of potentially high level infectivity

	Question to Patient	Notes to clinician
1	<p>Have you a history of CJD or other prion disease in your family? If yes, please specify.</p>	<p>Patients should be considered to be at risk from genetic forms of CJD if they have or have had:</p> <ul style="list-style-type: none"> i) Genetic testing, which has indicated that they are at significant risk of developing CJD or other prion disease; ii) A blood relative known to have a genetic mutation indicative of genetic CJD or other prion disease; iii) 2 or more blood relatives affected by CJD or other prion disease
2	<p>Have you ever received growth hormone or gonadotrophin treatment?</p> <p>If yes, please specify:</p> <ul style="list-style-type: none"> i) whether the hormone was derived from human pituitary glands ii) the year of treatment iii) whether the treatment was received in the UK or in another country 	<p>Recipients of hormone derived from human pituitary glands, e.g. growth hormone or gonadotrophin, have been identified as at increased risk of sporadic CJD.</p> <p>In the UK, the use of human-derived growth hormone was discontinued in 1985 but human-derived products may have continued to be used in other countries.</p> <p>In the UK, the use of human-derived gonadotrophin was discontinued in 1973 but may have continued in other countries after this time.</p>
3	<p>Have you ever had surgery on your brain or spinal cord?</p>	<p>(a) 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).</p> <p>(b) NICE guidance emphasises the need for a separate pool of new neuroendoscopes and reusable surgical instruments for high risk procedures on children born since 1st January 1997 and who have not previously undergone high risk procedures. These instruments and neuroendoscopes should not be used for patients born before 1st January 1997 or those who underwent high risk procedures using reusable instruments before the implementation of this guidance.</p>

J6. The actions to be taken following the patient's response to the above questions are:

Patient's response	Action
No to all questions	Surgery or neuro-endoscopy can proceed using normal infection prevention and control procedures.
Yes to any of questions 1, 2 or 3	<p>Further investigation into the nature of the patient's CJD risk should be undertaken, and the patient's CJD risk assessed. This assessment of CJD risk should be recorded in the patient's medical notes for future reference.</p> <p>If the patient is found to be at increased risk of CJD or vCJD following investigation, or the risk status is unknown at the time of the procedure, special infection prevention and control precautions should be taken for the patient's procedure including quarantining of instruments, and the local infection prevention and control team should be consulted for advice. Part 4 of this guidance provides advice for the precautions to be taken during the treatment of patients with or at increased risk of CJD or vCJD, and Annex F provides information on neuro-endoscopic procedures.</p> <p>If the patient is found to be at increased risk of CJD or vCJD they should also 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 Incidents Panel: http://www.hpa.org.uk/CJDIncidentsPanel</p> <p>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/</p> <p>Patients who are at increased risk of sporadic CJD due to receipt of human-derived growth hormone or gonadotrophin should be offered the opportunity of referral to the UCL Institute of Child Health, London. Contact: leahdavidson@msn.com, 020 7404 0536</p>
Unable to respond	See paragraphs J7 – J10 below for advice.

Emergency surgery or neuro-endoscopy which may involve contact with high risk tissue

- J7. In the event that a patient about to have emergency surgery or neuro-endoscopy is physically or otherwise unable to answer any questions, a family member, or someone close to the patient (in the case of a child, a person with parental responsibility), should be asked the CJD risk questions as set out in [Table J1](#) prior to the surgery or neuro-endoscopy.
- J8. If the family member, or someone close to the patient, is not able to provide a definitive answer to the CJD risk questions, the surgery or neuro-endoscopy should proceed but all instruments should be quarantined following the procedure (see [Annex E](#) of this guidance for details on quarantining). The patient's GP should be contacted after the surgery or neuro-endoscopy, and enquiries made as to whether the patient is at increased risk of CJD/vCJD according to the questions as set out in [Table J1](#).
- J9. The actions to be taken following the GP's response to the questions in [Table J1](#) are:

GP's response	Action
No to all questions	The instruments can be returned to routine use after undergoing normal decontamination processes.
Yes to any of questions 1, 2 or 3	<p>Further investigation into the nature of the patient's CJD risk should be undertaken, and the patient's CJD risk confirmed or rejected. Confirmation or rejection of CJD risk should be recorded in the patient's medical notes for future reference.</p> <p>If the patient is found to be at increased risk of CJD or vCJD following investigation then the quarantined instruments should be destroyed. Alternatively, instruments destined for disposal may instead be retained for research – refer to Annex E for details.</p> <p>The patient's GP should inform the patient that they are at increased risk of CJD or vCJD and provide them with further information and advice. This is available from: http://www.hpa.org.uk/CJDIncidentsPanel</p> <p>Patients who are at increased risk of genetic forms of CJD may benefit from discussions with the National Prion Clinic, based at the National Hospital for Neurology and Neurosurgery, Queen Square, London: http://www.nationalprionclinic.org/</p> <p>Patients who are at increased risk of sporadic CJD due to receipt of human derived growth hormone or gonadotrophin may benefit from discussions with the UCL Institute of Child Health, London. Contact: leahdavidson@msn.com, 020 7404 0536</p>
Uncertain about any of questions 1, 2 or 3	The instruments should be kept in quarantine. The local infection prevention and control team should carry out a risk assessment, and they may wish to involve the local Control of Communicable Disease Consultant in this process. The outcome of the risk assessment should determine whether or not to return the instruments to routine use.

Additional actions to be taken during pre-surgery assessment for CJD risk

- J10. In addition to asking the patient CJD/vCJD risk questions, the following actions should also be carried out before any surgical or endoscopic procedure involving contact with high risk tissue. The clinician undertaking the pre-surgery assessment should:
- Check the patient's medical notes and/ or referral letter for any mention of CJD or vCJD status
 - Consider whether there is a risk that the patient may be showing the early signs of CJD or vCJD, i.e. consider whether the patient may have an undiagnosed neurological disease involving cognitive impairment or ataxia
- J11. These actions, in conjunction with the CJD/vCJD risk questions, will minimise the chance of a CJD incident occurring and therefore reduce the risk of transmission of CJD or vCJD to subsequent patients.

Infection prevention and control guidance

J12. [Part 4](#) of this Guidance provides advice on the special infection prevention and control precautions that should be taken for patients with, or at increased risk of, CJD or vCJD, and [Annex F](#) provides information on endoscopic procedures.

Patients at increased risk of CJD or vCJD

J13. As outlined in Table 4A in [Part 4](#), a number of patients have been identified as at increased risk of CJD or vCJD on the recommendation of the CJD Incidents Panel. [Paragraphs J15 to J17](#) provide some further information on these individuals and the steps taken to ensure that health care staff are informed of their risk status.

J14. Patients identified to be at increased risk include:

Related to blood transfusions

- People who have received blood or blood components from someone who went on to develop vCJD
- People who have given blood or blood components to someone who went on to develop vCJD
- People who have received blood or blood components from someone who has also given blood or blood components to a patient who went to develop vCJD
- People who have received blood or blood components from 300 or more donors since 1990 (additional information on how this group is defined can be found in the ACDP TSE guidance FAQ document^[1]).

Related to surgery

- People who have had surgery using instruments that had been used on someone who developed CJD
- People who have had an intradural neurosurgical or intradural spinal procedure before August 1992
- People who have received an organ or tissue from a donor infected with CJD or at increased risk of CJD

Related to other medical care

- People who have been treated with certain UK sourced plasma products between 1990 and 2001
- People who have been treated with growth hormone sourced from humans (before 1985)
- People who have been treated with gonadotrophin sourced from humans (before 1973)
- People who have been told by a specialist that they have a risk of developing the genetic form of CJD

[1]

https://www.gov.uk/government/uploads/system/uploads/attachment_data/file/209775/Frequently_Asked_Questions.pdf

- J15. When someone is notified that they are at increased risk of CJD or vCJD, they are asked to take certain precautions to reduce the risk of spreading the infection to others. These include:
- Not donating blood, tissue or organs;
 - Informing healthcare staff if they need to undergo an invasive surgical, medical or dental procedure;
 - Informing a family member or someone close to them, in case they need emergency surgery or endoscopy in the future
- J16. The individual's GP is asked to record the patient's CJD risk status in their primary care records. The GP should also include this information in any referral letter should the patient require invasive surgical, medical or dental procedures.
- J17. Further information on the work of the CJD Incidents Panel is available on the HPA website: <http://www.hpa.org.uk/CJDIncidentsPanel>

Training

- J18. Health services should ensure that healthcare staff conducting pre-surgery assessments receive instruction and/or training necessary to understand the reasons for asking these questions. **It is important that these questions are asked in a manner that does not cause undue anxiety, and therefore the questioner should be prepared and able to reassure the patient, and provide further information if needed. Information for patients is available from the CJD Incidents Panel website at: <http://www.hpa.org.uk/CJDIncidentsPanel>**

Information for patients undergoing surgery or neuro-endoscopy on high risk tissues**Appendix B**

Part of your routine assessment before surgery includes some questions to find out whether you could have an increased risk of Creutzfeldt-Jakob disease (CJD). We will ask you:

Have you ever been notified that you are at risk of CJD or vCJD for public health purposes?

Have you any history of CJD or other prion disease in your family?

Have you ever received growth hormone or gonadotrophin treatment?

Have you had surgery on your brain or spinal cord at any time in the past?

What is CJD?

Creutzfeldt-Jakob disease (CJD) is a rare brain disorder that affects about 1 in a million people each year. CJD is thought to be caused by the build up in the brain of an abnormal form of a protein called a 'prion'. Unfortunately CJD is fatal, and as yet there is no known cure. There are different types of CJD, including variant CJD (vCJD). vCJD is caused by eating meat from cows infected with BSE.

How can CJD spread from person to person?

A person who is infected with CJD may have abnormal prion protein in their body for years before becoming ill. If that person has an operation, or donates blood, tissues or organs, during that time, the abnormal prion protein that causes CJD could spread to other patients.

Why are we asking you about CJD before your operation?

The abnormal prion protein that causes CJD is very hard to remove or destroy. If surgical instruments are used on a patient who is infected with CJD they may still have prion protein on them, even after they have been properly washed and disinfected. They could then spread CJD to other patients. This is particularly important for operations on the brain, spinal cord and the back of the eye as these parts of the body contain the largest amount of abnormal prion protein.

What have these questions got to do with CJD?

CJD has been spread in several ways and different groups of people may have an increased risk of CJD.

We ask whether there is anyone in your family who has had CJD because some types of CJD can be inherited. These types of CJD are caused by faulty genes and may be passed from parent to child.

We ask whether you have had surgery on the brain or spinal cord because some of these operations used to use grafts of 'dura mater' (the tough lining round the brain and spinal cord). Some of these grafts have been linked to CJD infection - these grafts are no longer used.

We ask whether you have been treated with growth hormone or gonadotrophin infertility treatment because these used to be prepared from pituitary glands. Some of these hormone treatments have been linked to CJD infection - these hormones are no longer used.

We ask whether you have had a large number of blood transfusions as this could be related to an increased risk of variant CJD (vCJD). vCJD is the type of CJD which is caused by eating meat from cows infected with BSE. vCJD can be spread through blood transfusions.

What happens if I answer 'Yes' to any of these questions?

If you answer 'Yes' to any of these questions, medical staff will now examine your medical records in more detail to determine whether or not you may have an increased risk of CJD.

What will happen then?

If you do have an increased risk of CJD special precautions will be taken with the surgical instruments used in your operation. Your GP will be informed and will ask you to come and discuss what this means in more detail.

Please remember that the overall risk of CJD spreading by these routes is generally **very low**. These questions are an extra measure to prevent CJD spreading through surgery. **This should not affect the medical care you receive now or in the future.**

What if I don't have a GP?

The health protection unit for your area will make sure that another doctor discusses this with you.

Can I have a blood test to see if I am infected with CJD?

Unfortunately there is no blood test available yet which could show if you have CJD.

Where can I find out more?

The following organisations offer further information and support.

- Public Health England: www.hpa.org.uk/cjd
- CJD Support Network website: www.cjdsupport.net
- National CJD Surveillance Unit website: www.cjd.ed.ac.uk
- National Prion Clinic website: www.nationalprionclinic.org/