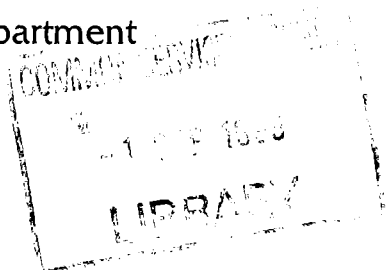




SCOTTISH EXECUTIVE

Health Department



Dear Colleague

VARIANT CREUTZFELT-JAKOB DISEASE (vCJD): *Minimising the risk of transmission*

Summary

This MEL is largely based on a Health Service Circular being issued by the Department of Health in England. It describes the present state of knowledge of the risks of transmission of variant Creutzfeldt-Jakob Disease (vCJD) from one patient to another. It details the action that health organisations and clinicians should already be taking to reduce the risk of transmission and recommends some further precautionary measures. A further MEL *NHS in Scotland: Decontamination Guidance*, will be issued shortly with an accompanying CD-ROM. When received, it should be read in conjunction with this document.

Action

Chief Executives of NHS Trusts and General Managers of Health Boards should take action to ensure that there are proper arrangements in place to comply with existing safeguards and new guidance as set out in this MEL. The key messages in this MEL are:

Existing safeguards:

- (i) All cases where CJD of any type is a possible diagnosis should be reported to the National CJD Surveillance Unit (CJDSU) so that any necessary action can be taken.

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- (ii) Effective and thorough cleaning of surgical instruments, to remove as much organic debris as possible, before sterilization, makes the major contribution to risk reduction. It is therefore essential that all existing cleaning and sterilization procedures operate to the highest standards, in line with extant guidance. Instruments designated for a single episode of use should be discarded after use and never reprocessed.
- (iii) The guidance produced jointly in April 1998 by the Advisory Committee on Dangerous Pathogens (ACDP) and the Spongiform Encephalopathy Advisory Committee (SEAC) on safe working and the prevention of infection must be followed to ensure optimum clinical care and management of patients with CJD of any type.

New advice

- (iv) Single-use kits should always be used for lumbar-punctures.
- (v) Where there are practical options for using single-use instruments, which do not compromise clinical outcome, consideration should be given to using these for surgical procedures.

Chief Executives of NHS Trusts should ensure that all relevant staff are fully conversant with the procedures set out in this MEL. In particular they should draw it to the attention of Medical Microbiologists, Infection Control Doctors, Infection Control Nurses and Operating Theatre Managers, Estates Managers and Sterile Service Managers.

General Managers of Health Boards should ensure that this MEL is drawn to the attention of Estates Managers and that copies are forwarded to all registered private, and voluntary, hospitals and clinics. They should also assure themselves that general practitioners, who undertake surgical procedures, and dental practitioners are fully aware of the contents of this MEL. From 1 October 1999 responsibility in respect of FHS contractors in mainland Health Boards will lie with Chief Executives of Primary Care Trusts.

Yours sincerely



Sir David Carter
Chief Medical Officer



Dr Kevin Woods
Director of Strategy and
Performance Management

THE EMERGENCE OF vCJD

Creutzfeldt-Jakob disease (CJD) in its classical form was first described in the 1920s. It is one of a group of diseases called transmissible spongiform encephalopathies (TSEs) which can occur in people or animals. The diseases are characterised by degeneration of the nervous system and are invariably fatal.

CJD in its classical form is the commonest of the human TSEs but it is still rare with an annual incidence across the world of 0.5 to 1.0 cases per million population. In Britain, there have been about 35 cases per year. The average age of onset of classical CJD is between 55 and 75 years. Classical CJD has no known cause in the majority of cases. However, about 15% of cases are inherited and are caused by gene mutations. About 1% of cases in the past resulted from transmission as a result of medical treatments such as injections of growth hormone derived from human pituitary glands, corneal transplants, or brain surgery which involved contaminated instruments.

Early in 1996, the National CJD Surveillance Unit, which is based in Edinburgh, identified a form of CJD that differed from previously recognised types of the disease. The patients affected were usually younger, their symptoms were different and the appearance of their brain tissue after death was not the same as in the classical form of CJD. The disease was initially labelled new variant CJD (nvCJD), and is now known as variant CJD (vCJD).

The number of definite and probable vCJD cases in the UK by the end of July 1999 is 43, all of whom have died. In 1998 16 people died of vCJD, compared to 10 in each of the previous two years. It is too soon to tell whether this increase in incidence will be sustained, or what the eventual number of vCJD cases will be.

The precise nature of the agent which causes vCJD is not known, but the most likely theory implicates an abnormal form of a protein which is called a "prion". Normal prion proteins are distributed throughout nature and are found in the tissues of healthy people and animals. It is believed that prions can cause disease when they become altered in shape, by folding in an abnormal way. The abnormally shaped prion protein then influences the normal form of the protein to alter its shape. This leads to destruction of nervous tissue, particularly in the brain, giving it a spongy appearance under the microscope.

The Government's Spongiform Encephalopathy Advisory Committee (SEAC) concluded that the most likely explanation for the emergence of vCJD, was that it had been transmitted to people as a result of the consumption of tissues from animals suffering from Bovine Spongiform Encephalopathy (BSE).

THE POSSIBILITY OF PERSON-TO-PERSON SPREAD

Available epidemiological evidence suggests that normal social or routine clinical contact with a patient suffering from any type of CJD, including vCJD, does not present a risk to healthcare workers, relatives and the community.

The possibility that vCJD might be spread from person-to-person in health care situations arises for a number of reasons:

- classical CJD has been transmitted from person to person by medical procedures,
- abnormal prion protein has been demonstrated in the lymphatic tissue (including tonsils) of patients with established vCJD,
- abnormal prion protein has been demonstrated in the appendix of a patient who subsequently developed vCJD,
- abnormal prion protein may not be inactivated by normal sterilization procedures.

The recent identification of the abnormal form of prion protein in the appendix removed from a man some months before he went on to develop clinical signs of vCJD¹, was the first time that the presence of abnormal prion protein had been detected in peripheral tissues before the onset of clinical disease. Furthermore, in another study² lymphoreticular tissues (tonsils, spleen and lymph nodes) from patients with neuropathologically confirmed vCJD were found to be positive for the abnormal prion protein. Tissues from patients with other types of CJD, and from control patients, did not test positive. In addition, strain typing of abnormal prion protein showed a consistent pattern in the tissues of patients with neuropathologically confirmed vCJD, and was of a type different from the strain found in brain tissue from patients with other types of CJD.

These findings raise two main issues. Firstly, there is a theoretical possibility of transmission of abnormal prion protein from lymphoreticular tissues of a pre-symptomatic patient undergoing certain surgical procedures to another patient. Secondly, they raise the possible use of tonsil biopsy as a diagnostic test and as a way of establishing the prevalence of vCJD through population studies, once the relevance of positive results to clinical disease is elucidated.

1. ACTION: REINFORCE EXISTING SAFEGUARDS AGAINST TRANSMISSION

The following section of the MEL restates measures already covered in extant guidance. Health organisations should ensure that these are being rigorously adhered to.

a. Notification of new cases of CJD to the CJD Surveillance Unit in Edinburgh.

Any patient suspected on clinical grounds of having vCJD, or any other type of CJD, should be referred to the CJD Surveillance Unit in Edinburgh. Not only is this required for epidemiological and surveillance purposes, but it is necessary as a control measure to guard against the theoretical risk of transmission of vCJD. Failure to notify promptly would prevent early action to trace and withdraw any blood donations which may have been made

¹ Hilton et al.,(1998) *Lancet* , **352**: 703-704

² Hill et al., (1999) *Lancet*, **353**: 183-189

by a sufferer from vCJD and could also prevent the institution of other control measures. Contact details for the CJD Surveillance Unit are:-

Professor R G Will
Director, National CJD Surveillance Unit
Western General Hospital
Crewe Road
Edinburgh EH4 2XU Tel: 0131 332 2117 Fax: 0131 343 1404

b. Properly clean and sterilize surgical instruments before re-use.

The abnormal protein associated with TSEs, including vCJD, is very resistant to all common methods of inactivation. Expert advice is that effective cleaning of surgical instruments prior to sterilization is of the utmost importance in reducing the risk of transmission of vCJD via surgical procedures. It is therefore essential that all existing cleaning and sterilization procedures operate to the highest standards.

Comprehensive guidance on decontamination will be available on the CD ROM *NHS in Scotland: Decontamination Guidance*, which will accompany a further MEL to be issued very shortly.

Decontamination equipment will work less efficiently on instruments which are difficult to clean and/or in poor condition. Consideration should therefore also be given to the condition of surgical instruments in use. Devices that cannot easily be cleaned should be identified and, where practicable and in a planned programme, replaced with versions that are easier to clean.

c. Handling instruments used on known or suspected CJD patients.

Policy on the use and disposal of surgical, diagnostic or other instruments involved in the care of patients who have or are suspected of having any type of CJD is set out in the guidance produced jointly by the Advisory Committee on Dangerous Pathogens (ACDP) and the Spongiform Encephalopathy Advisory Committee (SEAC)³. This Guidance was covered by MEL (1998)47, issued on 6 July 1998. Chief Executives and others leading health organisations should ensure that all relevant staff are fully conversant with the procedures set out in this guidance.

The ACDP/SEAC guidance makes it clear that:

- Instruments and equipment used in the care of patients with confirmed CJD of any type should not be re-used and should be disposed of by incineration;
- Instruments used on patients suspected of having CJD of any type should be quarantined pending confirmation of a diagnosis (and then should be destroyed by

³ "Transmissible Spongiform Encephalopathy Agents: Safe Working and the Prevention of Infection", April 1998. Full text available on:
<http://www.official-documents.co.uk/document/doh/spongifm/report.htm>

incineration unless a definitive alternative diagnosis is confirmed). Supplementary advice for those limited circumstances where quarantining of instruments may be appropriate is set out in Annex A to this MEL;

- Certain categories of patient should be regarded as presenting a potential risk (defined in the guidance as recipients of hormones derived from human pituitary glands, recipients of human dura mater grafts, and people with a family history of CJD);
- Single-use instruments and equipment should be used wherever possible in cases involving patients in a risk category as defined in the guidance;
- Instruments and equipment used in procedures involving brain, spinal cord or eyes carried out on a patient in a risk category as defined in the guidance should be destroyed by incineration.

d. Exposure of a health worker or research worker to a case of CJD

Action must be taken when clinical, laboratory or research workers are exposed to the infective agent associated with CJD of any type. One aspect of this is the requirement under the Control of Substances Hazardous to Health Regulations 1999 (COSHH) to keep a list of employees exposed to Hazard Group 3 or 4 biological agents. Annex B to this MEL sets out how this should be interpreted in respect of possible exposure to the agent of CJD (which is classified as a Group 3 biological agent), in the healthcare setting.

e. Advice on the care of patients

The following resources are available to health professionals dealing with cases of CJD:

- The National CJD Surveillance Unit in Edinburgh can provide advice on all clinical and neuropathological aspects of CJD. It can be contacted at:

Professor R G Will
Director, National CJD Surveillance Unit
Western General Hospital
Crewe Road
Edinburgh EH4 2XU

TEL: 0131 332 2117 Fax: 0131 343 1404

- The Prion Unit at St Mary's Hospital, London specialises in the care of patients suffering from CJD. It can be contacted at:

Kathryn Prout
Clinical Nurse Specialist
Prion Unit
Department of Neurology
St Mary's Hospital

Praed Street
London W2 1NY

TEL: 0171 886 6883

- The CJD Support Network is a voluntary organisation established to provide help and support for patients of all types of CJD and their families. The Network has undertaken a case co-ordination initiative aimed at facilitating the co-ordination of care for patients affected by all types of CJD, and gives advice on case co-ordination enabling cost effective care and ensuring appropriate responses to carers' needs. It can be contacted at:

Gillian Turner
National CJD Co-ordinator
CJD Support Network
Birchwood
Heath Top
Ashley Heath
Market Drayton, Shropshire TF9 4QR
TEL: 01630 673 993

- The Human BSE Foundation is a voluntary organisation run by families of vCJD patients aimed at helping relatives, friends and carers of vCJD patients by providing support, information and practical advice. It can be contacted at:

The Human BSE Foundation
Greenfields, Bath Road
Devizes
Wiltshire SN10 1QG

TEL (Helpline): 01380 720 033

2. ACTION : EXTENSION OF SAFEGUARDS AGAINST TRANSMISSION IN RESPONSE TO THEORETICAL RISK FROM vCJD

This section of the MEL sets out the new measures being implemented and recommended by the Departments of Health. Health organisations need to ensure that measures for which they are responsible are fully implemented.

a. Use of leucodepleted blood in transfusions.

There is considerable uncertainty about whether or not the infectious agent may be present in human blood and to what extent it would present any risk of transmission. The Government's main advisory committee on TSEs, the Spongiform Encephalopathy Advisory Committee (SEAC), advised in June 1998 that if there were any infectious agent present in human blood it would most likely be in the white cells. The Committee therefore recommended that as a precautionary measure the use of leucodepletion (removal of white

cells) for all blood destined for transfusion should be extended as soon as **practically** possible, whilst ensuring that there was no adverse effect on the donation and supply of blood. Action has been taken by the UK blood services to achieve this. As a result, all blood donated after 31 October 1999 and issued as red cells or platelets will have been leucodepleted. Clinicians should always ensure that the use of any blood or blood component is clearly indicated clinically.

b. Use of non-UK sourced plasma and blood products

There is also a hypothetical risk that vCJD may be transmissible by blood products. The Government's main advisory body on the safety of medicinal products, the Committee on Safety of Medicines (CSM), therefore advised in May 1998 that manufactured blood products should not be sourced from UK plasma until the possibility of transmission can be assessed and, if necessary, an effective screening test is made available. This advice has been implemented and all major blood products (eg Factor VIII, immunoglobulins and anti-D for Rhesus negative pregnant women) are now made from non-UK plasma.

c. Transplantation of Organs and Tissue Implants

There is a theoretical but unquantifiable risk of transmission of vCJD by organs or tissues, including bone grafts, transplanted from those incubating vCJD. Current procedures to minimise that risk should be in line with the Microbiological Safety of Blood and Tissues for Transplantation (MSBT) guidance⁴ issued in 1996 and the guidance contained in this MEL. The MSBT guidance is under review and revised guidance is expected later this year. Procedures should be reviewed again as soon as the revised guidance is available.

In the interim, potential donations should be rejected in the following circumstances:

- Recipients of pituitary derived hormones such as human growth hormone or gonadotrophins,
- People known or assumed to have had human dura mater implanted including:
 - People who have had brain surgery before August 1992,
 - People who have had an operation for a tumour or a cyst of the spine before August 1992,
- People diagnosed or suspected of suffering from CJD of any type, or with a family history of CJD,
- People with degenerative neurological diseases of unknown causation,
- Recipients of corneal transplants.

⁴"Guidance on the microbiological safety of human tissues and organs used in transplantation", MSBT, March 1996

Human dura mater should not be used in surgical procedures because of the high risk of transmission of CJD. The licence for products derived from human dura mater was withdrawn in 1992.

Until there is an effective screening test for vCJD, the decision on whether to use an organ from a particular donor, particularly in the case of life-preserving organ transplants, will depend on clinical need.

d. Single-use instruments

Single-use kits must always be used for lumbar-puncture procedures.

The ACDP/SEAC guidance (see 1c above) states that instruments should not be re-used once they have been used on patients with known or suspected CJD of any type. Even if single-use disposable items are not available, the instruments should under no circumstances be re-used. The same precautions are recommended for patients in a risk category for CJD as defined in the guidance where the clinical intervention involves brain, spinal cord or eye. For these patients, where other tissues are involved, the use of single-use instruments is encouraged.

In the light of uncertainty about the tissue-distribution of infectivity in vCJD, where there are practical options for using single-use instruments, which do not compromise clinical outcome, consideration should be given to using these for surgical procedures.

Devices designated for single episodes of use must not be re-used under any circumstances. As they are not designed to be reprocessed, it will be impossible to ensure they are decontaminated effectively.

Recent advice (via MDA advice note 1999(02)) is that contact lenses issued to patients for a trial wearing should not be re-used. SEAC's view is being sought on the possible risks posed by the re-use of special complex diagnostic lenses and other instruments used in ophthalmology departments and optical practices which come into contact with the eye. In the meantime such devices and instruments may continue to be used, but the greatest care should be taken in their cleaning and preparation for safe re-use.

The implications of vCJD for procedures involving the brain and spinal cord are also under review. Meanwhile the ACDP/SEAC guidelines on brain, spinal cord and eye surgery should be followed.

Further advice will be issued as new information becomes available.

e. Identification and tracing of flexible endoscopes

While systems for routine tracing of instruments in the event of a vCJD related incident are being developed, immediate action as set out below should be taken to enable rapid tracing of flexible endoscopes.

Flexible endoscopes are expensive pieces of equipment and if they have to be quarantined as a result of use in a possible case of vCJD and then subsequently destroyed, there is a large cost attached. In such an eventuality, it is vitally important to be able to identify the instrument with certainty; if the relevant instrument cannot be distinguished from identical ones in a pool, it may be necessary to quarantine and possibly destroy all the endoscopes in the pool. All endoscopes must therefore have a unique identifier and this, together with the identifying details of the patients concerned, should be recorded in the endoscopy suite, theatre or outpatient department on every occasion that the instrument is used.

A REMINDER OF THE EXPERT ADVISORY COMMITTEE STRUCTURE.

The main expert Committees which advise the Government (on a UK basis) and the NHS on these matters are:

- Spongiform Encephalopathy Advisory Committee (SEAC)
 - advises on the scientific aspects of Transmissible Spongiform Encephalopathies
- Advisory Committee on Dangerous Pathogens (ACDP)
 - advises on all aspects of hazards and risks to workers and others from exposure to pathogens
- Microbiology Advisory Committee (MAC)
 - advises on microbiological aspects of equipment used and intended to be used in the Health Service, including disinfection and sterilization practices
- Microbiological Safety of Blood and Tissues for Transplantation (MSBT)
 - advises on measures to ensure the microbiological safety of blood and tissues for transplantation.
- Committee on Safety of Medicines (CSM)
 - advises on the safety of new medicines and the adverse reaction reports on medicines already on the market.

CONCLUSION

While no cases of vCJD to date have been attributed to transmission by medical interventions, the emerging findings on the distribution of abnormal prion protein in the organs and tissues of vCJD patients mean that the precautionary principle must apply to minimise the risk of transmission.

Annex A

QUARANTINING OF SURGICAL INSTRUMENTS

Paragraph 4.28 of the ACDP/SEAC guidance on "Transmissible spongiform encephalopathy agents: Safe working and the prevention of infection" allows for instruments that have been used on a patient suspected of having CJD of any type to be quarantined pending a confirmation of diagnosis. Although it is not expected that this facility will need to be used widely, the following supplementary advice has been prepared for reference in those instances where such quarantining may be appropriate.

At the completion of a surgical procedure undertaken on a patient suspected of suffering from CJD of any type, single-use instruments should be separated and disposed of by incineration, re-usable instruments should be washed to remove gross soil. Care should be taken to avoid splashing and generating aerosols by holding instruments below the surface of the water in a sink into which water is running and draining out continuously. Instruments should not be held directly under a flowing tap as this is likely to generate splashes. Operatives should wear protective gloves and either a visor or goggles and care must be taken to avoid penetrating injuries.

Instruments should be placed in a disposable instrument tray and allowed to air dry. They should then be placed in an impervious rigid plastic container with a close fitting lid. The lid should be sealed with heavy-duty tape (e.g. autoclavable tape) and labelled with the patient's identification (i.e hospital number, name and date of birth), the surgical procedure in which the instruments were used and the name of the responsible person (e.g. the theatre superintendent). The sealed box should be stored indefinitely in a suitable designated place until the outcome of any further investigation is known. The instrument tray should be disposed of by incineration.

If the patient is confirmed as suffering from CJD of any type, the box and its contents should be incinerated without further examination. If an alternative, definitive diagnosis is confirmed, the instruments may be removed from the box by the responsible person (or a named deputy) and sent to the Sterile Service Department (SSD) for processing in the usual way.

Records must be kept of all decisions, and the SSD must be told of the decision before the instruments are sent for routine processing.

Prolonged autoclaving or supplemental disinfection is not necessary for instruments removed from quarantine, after investigations have revealed that the patient on which they had been used was not suffering from CJD of any type.

Further guidance will be issued as new information on vCJD emerges.

What to include in the list of employees

The list must indicate the type of work done and, where known, record any specific exposure, accident or incident (some of which may be reportable under RIDDOR). This may include staff who have performed one of the above procedures on a patient subsequently shown to have CJD of any type, unless the procedure was conducted before the patient was likely to have been incubating the disease.

The list must be kept for at least 40 years after the last known exposure. The list is not the same as a health record required for the purposes of health surveillance under COSHH or the Management of Health and Safety at Work Regulations, however, it must be available to any doctor appointed for health surveillance (eg the local occupational health physician). It must also be available to any employee specifically responsible for health and safety (such as a safety manager). Each employee must have access to the information which relates to him or her personally. In practice the list might best be kept with other transferable confidential information (e g information open to access by authorised individuals) in the staff member's occupational health record. Guidance on record keeping can be found in the Health Services Advisory Committee guidance "The management of occupational health services for healthcare staff" (HMSO 1993, ISBN 0-11-882127-X).

The ACDP/SEAC guidance also recommends including certain other information about the member of staff (eg full name and maiden name for women; NI number; date of birth and dates of employment). Although this is not a legal requirement, it should improve the value of the list in any future epidemiological surveillance.

Annex B

RECORDING STAFF INVOLVED IN THE CLINICAL CARE OF CJD PATIENTS

Part 2 of the ACDP/SEAC guidance on "Transmissible spongiform encephalopathy agents: Safe working and the prevention of infection" provides guidance on health and safety law in relation to workplace exposure to TSE agents, including the requirement under the Control of Substances Hazardous to Health Regulations 1999 (COSHH) to keep a list of employees exposed to hazard group 3 or 4 biological agents. This Annex sets out how this requirement should be interpreted with respect to exposure to CJD of any type in the healthcare setting.

Employers are required to keep a list of employees exposed to the agent of CJD of any type when there is a deliberate intention to work with the agent or, in cases of unintentional exposure, if a risk assessment shows there is a significant risk. The risk is deemed to be significant if more than basic hygiene measures are necessary to protect staff or if the control measures listed in COSHH are specifically applied.

The ACDP/SEAC guidance gives advice on when a list should be kept. It advises that in the majority of clinical situations there is no significant risk of exposure to CJD of any type. Staff working with such patients would not need to be listed as exposed to CJD (although their work with patients may be deemed to involve a "significant risk" from other Group 3 or 4 agents).

The decision to keep a list will depend on a local risk assessment. It is important to emphasise that the list is required where there is a likelihood of exposure, not simply when there has been a known incident or accident (although it should also include details of these). In this sense it is not the same as the requirement to report certain diseases and accidents to HSE under the Reporting of Injuries, Diseases and Dangerous Occurrences Regulations (RIDDOR).

For other types of work, the guidance recommends additional control measures for staff undertaking certain procedures involving patients with clinical symptoms of CJD of any type or those defined as at risk of familial or iatrogenic CJD by virtue of their clinical history. Such staff may therefore need to be listed as being potentially exposed to CJD. Examples of when this may be required are:

- all those involved in laboratory research work with the agent of CJD of any type,
- any staff performing invasive clinical procedures on patients suspected to be suffering from CJD of any type, particularly where there is a risk of exposure to central nervous or eye tissue or other tissues known to contain CJD infectivity,
- laboratory staff handling tissue specimens from such patients, in either routine or specialist neuropathology laboratories,
- staff undertaking post-mortem examinations of patients who have died of CJD of any type or where CJD of any type is suspected.