CREUTZFELDT-JAKOB DISEASE: GUIDANCE FOR HEALTHCARE WORKERS
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I. Background

Purpose of the guidance

1. This guidance is intended to guide healthcare professionals caring for patients with Creutzfeldt-Jakob disease and their families. It has been written in recognition of the fact that some features of CJD may make care provision more difficult. These include: the rarity of the disease and uncertainty about the diagnosis in the early stages; the combination of psychiatric and physical symptoms; the rapidity of progression of symptoms; and the high media profile surrounding the condition. Patients with CJD are initially cared for in a variety of settings, including psychiatric, neurology, geriatric, and psycho-geriatric units. Coordination of care has been a particular difficulty in the past.

2. This guidance covers the care of patients with known or strongly suspected CJD. It addresses patients’ and families’ care and information needs, including coordination of care. It complements advice on care planning in the document “CJD: Good practice guidelines for social service professionals” issued by the CJD Support Network in 1998 (see section V).

National sources of advice and support

3. Because of the rarity of the disease, many healthcare workers will have had little or no experience of dealing with it. The Department of Health is therefore funding the following national sources of advice and support for professionals and carers:

• The National CJD Surveillance Unit (CJDSU) in Edinburgh was established in 1990 to carry out epidemiological surveillance of all types of CJD in the UK. In response to notification of a suspected case of CJD, a research registrar from the Unit will visit to examine the patient and interview the patient and family. The CJDSU does not undertake the management of patients but can offer further advice the investigation of patients with possible or suspected CJD, or medical management of symptoms. A National Care Co-ordinator is employed at the CJDSU to provide specialist expertise in CJD and act as an information resource for carers and professionals.

• In addition, The CJD Support Network has undertaken a case-co-ordination initiative aimed at facilitating the co-ordination of care of patients with CJD, and particularly ensuring that families’ needs and wishes are taken into account.

4. Full details of these are set out in section II. Contact details for these and other sources are summarised in Section V.
Aetiology

5. CJD refers to human spongiform encephalopathies, degenerative brain diseases which are invariably fatal. They cause characteristic microscopic spongiform changes in the brain on pathological examination.

6. The causative agent is remarkably resistant to conventional sterilisation and disinfection techniques. It is thought that CJD is caused by infectious proteins known as ‘prions’, which are rogue forms of a normal protein found in the brain.

7. CJD has a long incubation period, which is known to be up to 25 years or more in some types of the disease. There is no simple diagnostic test at present. The diagnosis can only be confirmed on neuro-pathological examination, by brain biopsy or post mortem.

8. CJD is not transmissible from person to person by normal contact.

Types of CJD

9. There are different types of CJD. These are:

   • **Sporadic, or classical, CJD**: This is currently the most common form. There are around 50 cases of classical CJD in the UK each year. It is very unusual in people aged under 40 years. The cause of classical CJD is unknown.

   • **Variant CJD**: This form has recently been recognised. At the time of writing there have been around 50 cases of vCJD in the UK in total. It affects younger people than classical CJD. The duration of illness is longer, lasting a median of 14 months. In the early stages, patients often present with personality change and psychiatric symptoms such as depression or withdrawal. It is thought to be causally related to exposure to the agent causing Bovine Spongiform Encephalopathy in cattle.

   • **Iatrogenic CJD**: Cases of CJD have been associated with treatments administered in the 1970s using human pituitary derived growth hormone, and with human dura mater grafts. A few cases have been reported associated with corneal grafts, and a few with contaminated instruments used in brain surgery.

   • **Familial prion disease**: There are other rare types of CJD that are familial. About two dozen families in the UK are known to be affected. The condition follows an autosomal dominant pattern of inheritance.

Clinical features

10. CJD is invariably fatal. The illness usually has a short duration after the onset of progressive symptoms but varies according to the type of CJD. The median illness duration is approximately 3-4 months in classical CJD, 14 months in vCJD and 2-
5 years in inherited forms. Clinical features vary depending on the regions of the brain affected but all patients experience very rapid deterioration.

11. The common features include:

- Personality change
- Psychiatric symptoms
- Cognitive impairment
- Neurological deficits, including sensory and motor impairments and ataxia
- Myoclonic jerks, or, less frequently, chorea or dystonia
- Rapid, or unpredictable stepwise, deterioration
- Increasing difficulty with communication, mobility, swallowing and continence
- Coma
- Death

Uncertainties about the diagnosis

12. Because of the rarity of the disease and the lack of a simple diagnostic test it is often difficult to confirm the diagnosis. Older patients with sporadic CJD may initially be given a diagnosis simply of dementia. Younger patients with variant CJD have often initially been given a diagnosis of depression. It should be stressed that many patients may benefit from skilled psychiatric management. However many patients’ families are angry that their relatives have been given a psychiatric diagnosis and managed in a psychiatric setting. This has sometimes proved a barrier in later communication with the family and is another factor making coordination of care more difficult. Professionals should be aware of these issues and acknowledge families’ feelings.

Diagnostic process

13. Patients and families should be given a clear explanation of all investigations in lay language, including what the investigations involve, the results and their implications.

14. Most patients become cognitively impaired and it is necessary to gain consent for investigations from a next of kin.

15. When the diagnosis of CJD is suspected, a number of investigations are usually carried out. These are helpful to exclude other conditions, and some findings support a diagnosis of CJD. The investigations commonly performed include:

- **Lumbar puncture:** It is not possible to detect prion protein in CSF. Recently a test for a protein in the CSF known as ‘14-3-3’ protein has been described. Detection of this protein supports the diagnosis of CJD.

- **EEG:** There are characteristic EEG abnormalities that are often seen in sporadic CJD. These are not seen in vCJD, although the EEG often becomes non-specifically abnormal.
• **CT scan:** This usually does not show any abnormalities in CJD but may exclude other conditions.

• **MRI scan:** This may show heightened signal in the caudate and putamen regions of the basal ganglia in sporadic CJD and high signal in the putamen in variant CJD. The presence of these abnormalities supports a diagnosis of CJD.

• **Tonsil biopsy:** Lymphoreticular tissues from patients with neuropathologically confirmed vCJD have been found to be positive for the abnormal protein associated with prion diseases.

• **Brain biopsy:** The only way to diagnose CJD with certainty is by neuropathological examination. Brain biopsy is carried out in some cases, but it is an invasive procedure and may give a negative result if an unaffected part of the brain is biopsied.

• **Genetic testing:** The genes responsible for familial types of CJD have been identified and are detectable on blood testing.

16. In familial forms of the disease, it is possible for family members to be tested to find out if they are likely to develop the disease. This predictive testing should only be carried out with full informed consent following specialist genetic counselling at a regional genetics centre. The leaflet ‘Prion disease’ produced by the CJD Support Network has information on familial CJD for families (see section V for details).
II. Care requirements and co-ordination

17. Patients with terminal or degenerative conditions benefit from coordination of care and the identification of a key worker. This guidance is for care of patients with CJD, but describes good practice that should be applicable to those caring for patients with a wide range of conditions. Further relevant advice on care planning for patients with CJD is available in the document ‘CJD: Good practice guidelines for social services professionals’ (see section V for details).

Health Act Partnership Arrangements

18. In putting together the most responsive arrangements for the patient, it may be worth considering the use of the Health Act Partnership Arrangements. Pooled funds, lead commissioning and integrated provision can be established for individual patients if that is necessary. In the case of CJD, it may be helpful for local partners to consider deciding in advance of a case in their area, to set up a pooled fund, which can be activated when necessary. Alternatively, a lead commissioner of all the services likely to be required could be identified. For more information on the partnership arrangements, including the money transfer powers, please see www.doh.gov.uk/jointunit/index.htm The powers are discretionary, and enable more customer focused and timely responses to be developed.

Support and advice

19. Because of the rarity and special features of the disease, local care services will need specialist support and advice. There are now two complementary sources of this support and advice that work in partnership with each other.

20. From January 2000 the National CJD Surveillance Unit (CJDSU) has employed a national care co-ordinator to provide specialist expertise in CJD and act as an information resource for carers and professionals. Following initial referral of a suspected CJD patient to the CJDSU for investigation, the co-ordinator will make contact and meet with the patient, their carers and professionals to discuss and help implement plans of care. He will keep in touch with patients, carers and professionals and monitor the response of local agencies. The co-ordinator is Gordon McLean and he can be contacted at the CJDSU on 0131 537 2129.

21. From April 1999 to March 2002 the Department of Health has funded a national CJD case co-ordination project run by the CJD Support Network. Mrs Gillian Turner, the case co-ordinator, will attend the first case conference of a patient with CJD by invitation and assist all those present by providing information on effective forms of support, encouraging co-ordination and partnership between services and applying a unique understanding of carers needs. She can be contacted on 01630 673 993.

22. Both co-ordinators will provide detailed advice to local agencies on the likely progression of the disease and the range of care services that may need to be put in place. They will emphasise the importance of appointing a key worker at the earliest stage to co-ordinate a speedy response, tailored to meet the needs of the patients and their families.
Identification of a key worker

23. All patients with a clinical diagnosis of CJD should have a named key worker identified to co-ordinate care for patient and family, regardless of the setting in which care is given.

24. The key worker should be identified as soon as possible after the diagnosis of CJD is considered likely. In practical terms, this usually means during the hospital admission in which CJD is diagnosed. There should be a case conference before the patient leaves hospital at which the key worker is appointed and initial arrangements made for the patient’s future care.

25. Where patients are discharged home, the key worker should be appointed, and a care package arranged, before discharge. This will ensure a smooth transition of care and support for relatives at a very vulnerable time. In the event that a patient goes home before the case conference, it is essential to ensure that the GP is aware of the situation and that all possible support is arranged. As part of discharge planning the needs of carers should also be assessed and they should have a copy of the care plan.

26. The key worker should be a professional with knowledge of local health and social services. Their discipline will depend on local circumstances, but could be social work, nursing, or a profession allied to medicine. He/she must be able to develop a good relationship with the patient and family and be allocated sufficient time to devote to them.

27. Where patients are to be cared for in hospital, nursing home or hospice, patients will have a named nurse, but a separate key worker should also be appointed. It is essential that the key worker is able to provide continuing support and coordination of care across all the settings in which the patient is cared. He/she also has an important role in providing support to the family as well as the patient, continuing after the patient has died. For these reasons, the key worker should be separate from, but work closely with, the named nurse.
**Role of the key worker**

28. The key worker role includes:

- Coordinating assessment and reassessment of patients’ care needs
- Coordinating assessment of the family’s/carer’s needs for other practical help and childcare
- Coordination of professionals providing care
- Advocacy for resources to meet the needs identified
- Ensuring flexibility of the care package, and availability of respite care for both patient and family
- Providing or arranging advice on financial help and benefits
- Providing information about the condition to the family/carer, as it develops
- Providing emotional support to the family/carer
- Providing information and helping the family with end of life decisions
- Helping with funeral arrangements
- Continuing emotional support after the patient dies, and assessment of need for formal counselling

**Coordination of care plan**

29. Whatever the discipline of the key worker, it is important that both health and social services professionals are involved in the care of patients. Regular case conferences involving all the relevant agencies should be held to plan and coordinate care of the patient. The key worker should be responsible for arranging these, and for the overall coordination of care.

30. As a patient’s condition deteriorates, needs often change rapidly. Patients with suspected CJD and their family/carer should have their needs frequently reassessed. The family/carer should be told how to contact the key worker between planned visits in the event of a change in the patient’s condition.

**Care needs**

31. The needs of patients with CJD, and their families, will vary and it is not possible to define a standard package that will suit all cases. It is most important that key workers are sensitive to patients’ changing needs, anticipate likely progression where possible and support families who wish to care for patients themselves. This means that a flexible and creative approach is required, responding the needs and wishes of each patient and family.

32. The following services should be considered when planning the care:

- Nursing care
- Social services input
- Advice on benefit entitlements
- Occupational therapy
- Dietetics
33. As the condition progresses, patients are likely to need help with:

- Washing and dressing
- Mobility
- Continence
- Feeding
- Communication

34. Professionals should be alert to the rapidity of deterioration that occurs in CJD and reassess patients’ care needs frequently.

35. Some patients have psychotic symptoms including hallucinations or delusions. Many develop a fear of strangers. Both families and professionals should be made aware that challenging behaviour, for example aggression or sexual disinhibition, may occur. These symptoms may pose a particular difficulty on acute medical wards. Where patients are disturbed, a registered mental nurse, community psychiatric nurse or registered mental handicap nurse should be involved in their care.

Continuity of carers

36. Some patients develop a fear of strangers. They are distressed if there are many, frequently changing, staff involved in their care. Care should be taken to achieve as much continuity of care staff as possible.

Involvement of the patient’s family

37. Many families find it is important for them to provide much of the care themselves. Regardless of the setting, they should be helped to do as much of the caring as they wish. They may require support to allow them to give time to caring for the patient, as well as help with hands on care of the patient.

38. Families often have great insight into patients’ changing abilities. Their opinions about these should be central to assessment of patients’ needs.

39. Families and carers should be involved in case conferences and should have a copy of the care plan, with contact numbers of the key worker.
Terminal care

40. The decision about the best place for patients to receive terminal care will depend on individual circumstances and involve patients’ families. In most cases, acute neurology or psychiatric units cannot provide the appropriate environment for longer term care of patients, so they should be transferred as soon as possible. Specialist palliative care services should be involved wherever possible.

41. Terminal care should be provided as near as possible to patients’ homes. Patients should be cared for in an environment appropriate to their age. This is especially pertinent for younger patients. The key worker should identify suitable places and help the family to choose the most suitable for their own circumstances. The CJD Support Network can provide help in finding a suitable place.

42. Patients develop difficulties swallowing and may aspirate food. Most are eventually unable to sustain adequate nutrition and the decision has to be made whether to introduce naso-gastric or gastrostomy feeding. Informed staff, with experience of these issues, should discuss end of life decisions, including feeding, with families in advance and in the light of the poor prognosis.

Support for families caring for patients at home

43. Families often feel overburdened by the large number of professionals involved. It is important that professionals are sensitive to this and minimise the number of different people coming into their homes.

44. Families caring for patients at home should be offered advice on practical care of patients, including movement and handling, and the management of challenging symptoms.

45. The rapidity of progression of CJD means that any necessary aids or equipment, including incontinence pads, should be provided rapidly when they become needed.

46. Many families would benefit from the provision of respite care and night nursing services. These should be provided promptly when needed.

47. As carers, families have a right to have their own needs assessed and any necessary help provided.

Other support for families

48. Families should be offered practical help with household tasks and childcare as well as help with care of the patient. This applies regardless of the setting in which patients are cared for. However, depending on their financial circumstances, families may be charged for these services.
49. Many families suffer financially when caring for a patient with CJD. They should receive information on benefits.

50. All families need a high level of emotional support, during and after the illness. Professionals providing care of patients should provide informal support, continuing after the patient dies.

51. The key worker should also make sure each family member has access to formal counselling, especially after the death. Some families do not find formal counselling helpful during the time they are caring for dying patients, but do find it beneficial after the patient has died. The key worker should remain available to families after the patients’ death and ensure they have access to counselling at the time it is needed.

**Support for professionals**

52. Professionals caring for patients are also often distressed by the experience. They should be offered support and counselling if required.

**Maintaining confidentiality**

53. Professionals involved in the care of patients should be aware of the media interest in CJD. It is important to take precautions to maintain patient confidentiality. For example, a code number rather than a name could be used on clinical samples. No information should be given about patients over the telephone until the caller’s identity has been verified. Patients’ names should not be displayed prominently on the ward.

**Handling the media**

54. Some families of patients with CJD have been subject to unwanted media attention. Professionals should be aware of this. In the case of vCJD, the Human BSE Foundation may offer advice to families on dealing with the media.

**Funeral arrangements**

55. Families often find it helpful to start making funeral arrangements before the patient dies. This gives them more time to make the arrangements according to their wishes. The key worker should discuss these issues with the family sensitively, before the death. Undertakers may be reluctant to handle cases of CJD or may apply unnecessary restrictions due to a perceived risk of transmission. These problems may be resolved if anticipated and discussed with them beforehand. If necessary the key worker may liaise with the undertaker on behalf of the family. (See sections 80-82 for advice on infection control for undertakers).
III. Information needs and communication

56. Communication between professionals and patients’ families can be difficult because of the uncertainties over the diagnosis and prognosis, and the extremely distressing nature of the condition. Professionals caring for patients should be aware of these problems and be as sensitive as possible in communication with patients and families.

57. It is important to record what families have been told about the diagnosis and prognosis. Care should be taken that families do not receive conflicting information from different professionals.

Communicating the diagnosis

58. Where possible, families should be told the diagnosis by a doctor with whom they have already established a rapport. A nurse or other suitable person should be involved, in the discussion and afterwards, to provide support. No member of the family should be told when they are alone. In line with good practice on breaking bad news, they should be offered support, time to ask questions and a later interview to discuss it further. This should be planned in advance so that support can be available, and they can be accompanied home if necessary.

59. Patients’ GPs should be kept informed of the patient’s condition and care arrangements. GPs have a role in terminal care, and in continued support of the family.

Likely disease progression

60. Families should be given information about the condition, in a proactive way, as the condition develops. The key worker should act as a point of contact to answer questions at any stage.

61. The features and order in which they develop will vary between patients. But families should be prepared for:

- Challenging behaviour and sometimes aggression
- Increasing mobility problems
- Incontinence
- Difficulty swallowing and drooling
- Inability to communicate
- Inability to recognise people
- Cortical blindness
- Coma and death
Information about post mortem

62. The possibility of post mortem should be discussed with the patient’s next of kin before the death by the clinician responsible for care, in a sensitive manner. Where possible the key worker should be present at such discussions. This should include the likelihood of brain and other tissues being removed for later examination and research. Explicit, informed consent should be sought for post mortem and for removal of tissues after death. Relatives should be informed that post mortem is not compulsory but is needed to confirm the diagnosis.
IV. Precautions, risks and infection control

63. Detailed guidance on infection control of CJD in a healthcare setting is included in the document ‘Transmissible spongiform encephalopathy agents: safe working and the prevention of infection’. A Health Service Circular ‘Variant CJD: minimising the risk of transmission’ has also recently been published. These documents should be consulted for detailed advice, including advice on infection control for invasive clinical procedures (see section V for details).

64. The guidance identifies three patient risk groups. The following points are summarised from the guidance on care of patients with known or suspected CJD.

65. In familial types of CJD only, some precautions are necessary for invasive procedures on close family members of patients who are at risk of CJD. Detailed advice on this is available in the guidance.

66. Current evidence suggests that normal social or routine clinical contact does not present a risk to healthcare workers, families or others.

67. Special precautions are only required for handling high risk tissues. These are central nervous or eye tissue, including cerebro-spinal fluid.

68. Patients with CJD may be nursed on an open ward or at home, with no special precautions other than the standard good infection control practice that would apply to any other patient. Local infection control policies should be consulted for more detailed information on universal infection control precautions.

Universal precautions in hospital

69. Universal infection control precautions should be applied as they are for all other patients to safeguard the well being of the patient and the carer. This includes the need for handwashing before and after any procedure and the use of gloves and aprons when body fluids are involved.

70. There is no evidence of infectivity in saliva, body secretions or excreta. Used or fouled bed linen should be washed and dried in the usual way and in accordance with current guidance. Gloves should be worn and hands washed and dried after contact but no other precautions are necessary.

71. Should sheets become contaminated with cerebrospinal fluid or other high risk material they should be disposed of by incineration.

Clinical procedures

72. There is uncertainty about the risk of transmission from blood, but universal infection control should minimise any risk that may exist. Care should be taken
when dealing with sharps and needles should never be re-sheathed. These precautions should apply as for any other patient.

73. Any accident involving sharps or contamination of abrasions with blood or body fluid should be encouraged to bleed under a running tap for several minutes, washed gently with soapy water, rinsed, dried and covered with a waterproof dressing. The incident should be recorded and reported to senior managers.

74. Blood spillages and spillage of high risks body fluids. Wearing protective clothing (gloves and plastic apron), cover area with paper towels to absorb fluid. Then cover area (including towels) with either 10,000 ppm hypochlorite solution (1%) eg. Milton or sprinkle with Na DCC granules (Sodium dichloroisocyanurates) eg. Precept and leave for 2-3 minutes. Dispose of all waste into yellow clinical bags and incinerate. Then clean the area with detergent and warm water and dry.

75. For all other body fluid spillage eg. Urine/vomit/faeces, absorb fluid with paper towels, as above. Dispose of waste into yellow clinical bags and incinerate. Clean the area with detergent and warm water and dry. The application of a disinfectant solution is unnecessary.

76. All clinical waste materials should be disposed of by incineration. For the safe handling of infective material double bagging is advisable.

77. Routine clinical specimens should be stored and transported as local policy dictates. It is important to ensure that all specimens are clearly labelled and securely bagged for transportation.

78. The guidance should be consulted for advice on the precautions to be taken for any invasive procedure, including lumbar puncture, and handling of high risk specimens.

**Universal precautions in the home setting**

79. Although CJD is not thought to present a risk through normal social or routine clinical contact, families caring for patients at home should be advised of the standard infection control practice that would apply to any patient. They should be provided with gloves, paper towels, bags and sharps containers as appropriate.

80. Families should not be dissuaded from ordinary contact with patients but should wear gloves and aprons if handling body fluids.

81. In the home setting, patients’ clothes and bed linen may be washed as normal, but fouled linen should not be washed with other laundry. Where patients are incontinent, a laundry service can be of great help to carers.

82. Families should be provided with plastic bags for any clinical waste materials and sharps containers if appropriate. Provision should be made for the removal of clinical waste and sharps from the home for incineration.
83. Spillages of body fluids, including blood, should be removed using absorbent towels (e.g. kitchen paper) and the surface washed thoroughly with detergent and warm water. Disposable gloves and apron should be worn. Health professionals involved in caring for the patient should be in contact with the local authority to ensure that appropriate arrangements for the removal and disposal of waste are put in place.

After death

84. After death, the body should be placed in a body bag and removed to the mortuary using universal infection control measures.

85. If post mortem has not been carried out, there is no need for additional precautions other than those applied for any other patient. A theoretical risk of contamination with infectious material arises after post mortem examination in which the cranium has been opened. For this reason, contact with the body should be minimised following post mortem.

86. The guidance should be consulted for advice on precautions to be applied during post mortem examination.

87. Undertakers should use the general precautions required for handling intact bodies. Following post mortem examination it is advisable to minimise contact, particularly in circumstances where penetrating injuries could arise.

88. It is advisable to avoid embalming.

89. Relatives need not be discouraged from viewing the body or from superficial contact such as touching the face.

90. There is no need to discourage burial and no need for extra precautions for either burial or cremation.
V. Where to get help and advice

National CJD Surveillance Unit

Professor R G Will
National CJD Surveillance Unit
Western General Hospital
Crewe Rd
Edinburgh EH4 2XUT

Tel: 0131 332 2117
Fax: 0131 343 1404

National Care Co-ordinator for CJD

Gordon McLean can be contacted at the CJD SU on 0131 537 2129.

CJD Support Network

Gillian Turner
National CJD Co-ordinator
CJD Support Network
Birchwood
Heath Top
Ashley Heath
Market Drayton
Shropshire TF9 4QR

Tel: 01630 673 993

The Prion Unit

The Prion Unit at St Mary’s Hospital, London specialises in the care of patients suffering from all forms of CJD. It provides assessment, diagnosis and ongoing support for patients suffering from CJD.

Mr Colm Treacy
National Prion Clinic
Department of Neurology
Cambridge Wing
St Mary’s Hospital
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Human BSE Foundation

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.

The Human BSE Foundation
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Bibliography


