



## CREUTZFELDT-JAKOB DISEASE:

*Guidance for Health and  
Social Care Workers*

Department of  
Health, Social Services  
and Public Safety

An Roinn Sláinte,  
Serbhísí Sóisialta agus  
Sabháilteacta Poiblí

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## FOREWORD

Creutzfeldt-Jakob Disease, although rare, is always fatal and can have devastating effects on the patient, their family and carers. This guidance, which is timely due to the recent reported increase in the incidence of variant Creutzfeldt-Jakob Disease in the UK, should assist Health and Social Care Services to respond rapidly in a co-ordinated manner to the physical, social and psychological needs of the patient and their carers as these arise. I therefore recommend this guidance to Health and Social Services professionals as a resource to promote provision of a quality service.

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## I. INTRODUCTION

### a. *Background*

#### *Purpose of the guidance*

1. This guidance is intended to guide local health and social care professionals in caring for both patients with Creutzfeldt-Jakob disease (CJD) and their families. Certain features of CJD may make care provision more difficult and therefore a working group (membership in Annex B) was set up by the DHSS&PS to produce guidance. These features 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. The National CJD Surveillance Unit in Edinburgh does not undertake the management of patients but can offer further advice on these matters.

#### *Sources of advice*

3. There are several organisations and individuals that can advise those caring for patients with CJD. These organisations and are listed and described in Section V. Useful references are also listed.

## ***Aetiology***

4. 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.
5. 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.
6. 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.
7. CJD is not transmissible from person to person by normal contact.

## ***Types of CJD***

8. 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.
  - **Variante CJD:** This form has recently been recognised. At the time of writing there have been around 80 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.

## ***b. Clinical features***

9. 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.
10. 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***

11. 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.

### ***c. Diagnostic process***

12. Patients, due to their illness, may find investigations physically difficult, frightening, or may not understand what is required of them or why they should co-operate with staff during the investigation. It is important that patients and families should be given a clear explanation in lay language and as often as required of all investigations, including what the investigations involve, the results and their implications.

13. To facilitate the diagnostic process, where appropriate, a member of the patient's family, or a carer, should be allowed to accompany the patient during investigations.
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 Cerebro Spinal Fluid (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, but will not identify every case and can be present in other conditions.
  - **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 neuro-pathological 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).

#### ***d. Planning for future demands***

17. The DHSS&PS will monitor the number of cases of CJD within Northern Ireland and ensure each Health and Social Services Board has an appropriate strategy in place to provide the necessary services.
18. Health and Social Services Boards should ensure Trusts have drawn up contingency plans (as it is possible that the number of cases could increase considerably) and that each Trust has appointed a Designated Senior Officer to support staff with the management of CJD patients.
19. Trusts need contingency plans available and for plans to be effective, finance and resources are an essential requirement. Community care plans should take account of the effect of CJD in

service development and the need for resources in planning budgetary requirements.

- 20.** Each Trust should identify a Designated Senior Officer as a contact person for both staff within the Trust and also other organisations. The Designated Senior Officer should be at a senior management level and able to co-ordinate services to ensure relevant guidance is followed and the patient, family and carers' needs are met.
  
- 21.** It should be recognised that training is often required for professionals who have gaps in skills, knowledge and understanding of the disease. Workers need to know the disease and understand its implications, particularly the rapidity of its progression. There needs to be an awareness of the trauma of families, affected by the disease, who see the rapid decline of loved ones. Safety issues for patient and carers are paramount and, with rapid onset of the disease, safety needs to be high on the agenda.

## II. CARE REQUIREMENTS AND CO-ORDINATION

22. To ensure local awareness of up to date best practice in patient investigation and management there is a requirement for a local Regional Expert to be identified. This person, through maintaining links with national organisations and experts, would be able to provide advice to local professionals on case management, including any recent advances. The Local Regional Expert is Dr Tom Esmonde and he can be contacted at the Department of Neurology, Royal Victoria Hospital on 028 9089 4921.
23. Patients with life shortening or degenerative conditions benefit from coordination of care and the identification of a key worker. This guidance is for the care of patients with CJD, but describes good practice that should be applied to those caring for patients with a wide range of conditions.

### *Identification of a key worker*

24. All patients with a clinical diagnosis of CJD should have a named key worker identified to coordinate care for patient and family, regardless of the setting in which care is given. There should be close linkage between primary and secondary care.
25. The key worker should be identified as soon as possible after a 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 to take forward the arrangements made for the patient's care from that moment onwards. The National CJD Co-ordinator from the CJD

Support Network (Mrs Gillian Turner) should be invited to the initial case conference and can be contacted on 01630 673993

- 26.** Where patients are discharged home a care package should be 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.
- 27.** 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, medical 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.
- 28.** 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 for. He/she also has an important role in providing support to the family as well as the patient, which may continue after the patient has died. For these reasons, the key worker should be separate from, but work closely with, the named nurse.
- 29.** 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.

30. 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.
31. 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.

### ***Role of the key worker***

32. 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 and 24 hour community 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*

33. 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 and support for carers. The key worker should be responsible for arranging these, and for the overall coordination of care. Funding of the care plan should be agreed at the case conference. A model of some of the issues to be covered in case conferences is included in Annex A.
34. 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 need to respond quickly is vital due to the sudden deterioration of most people suffering from the disease, who within a matter of a few months, can move from being a healthy person to death. **The need for a quick response cannot be emphasised enough and this should be facilitated by anticipating future needs before they arise.** Timing is crucial

and all agencies must appreciate the importance of prompt action. Social services must be aware of need for initial urgent response, eg same or next day. They should consider if a ‘fast track’ approach to avoid bureaucratic delays is required on occasions, eg if there is a request for an orange badge and the need is obvious. 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.

35. People living alone in some respects are more vulnerable and will certainly need a care plan that takes account of their own individual circumstances. However, it may be dangerous for them to continue living alone, and it may also be appropriate to consider the need for an advocate to act on their behalf.
36. Some people, for example the homeless, could fall through ‘the gap’ in services. There is concern from the CJD Support Network in relation to this group if they have contracted the disease.
37. The minimum standards that people with CJD and carers can expect are:
  - **Assessment**
  - **Care plan**
  - **Implementation**

All with a sense of purpose and urgency.

### ***Care needs***

38. 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 to the needs and wishes of each patient and family.

39. Those who care for people have a right to an assessment in their own right. It is essential that professionals do not neglect or forget the needs of carers who are often providing 24 hour care, seven days a week. A husband/wife must be given the support/respite to fulfil other family roles.
  
40. The following services should be considered when planning the care:
  - Nursing care
  - Social services input
  - Advice on benefit entitlements
  - Occupational therapy
  - Dietetics
  - Physiotherapy
  - Speech and Language therapy
  - Laundry facilities
  - Specialist continence advice & services
  - Specialist palliative care
  - Voluntary sector services, eg Crossroads Caring for Carers (N.I.) Ltd, Hospice etc.
  
41. As the condition progresses, patients are likely to need help with:
  - Washing and dressing
  - Mobility
  - Continence
  - Feeding
  - Communication

42. Services must be appropriate to each individual. One concern of carers and service users is that age related services, eg young adult services, are often lacking. Whether appropriate services will be provided is a worry, and it is devastating for carers to see people receiving care in a setting inappropriate for their age group.
43. The services that will be required depend on specific needs. Often daily help and personal care are requested. Sitting services are also vital in order to give support and respite to carers.

### ***Involvement of the patient's family***

44. 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. A frequent concern and criticism from service users and carers is that professionals insist on things being done their way. Many people have their own individual ways of doing things and professionals need to be sensitive and ask people how they wish the service to be delivered.
45. Families often have great insight into patients' changing abilities. Their opinions about these should be central to assessment of patients' needs.
46. 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.

### *Continuity of carers*

47. Some patients develop a fear of strangers. They are distressed if there are many, frequently changing, staff involved in their care. As far as possible there should be continuity of care staff.
48. As the patient's condition deteriorates there will often be a requirement for extensive and possibly 24 hour nursing care. Although it is usual for a variety of statutory and voluntary agencies to provide out-of-hours nursing care in the patient's home, nevertheless, for this group of patients the key worker should ensure that the minimum number of professionals required should provide care. These professionals would ideally be already known to the patient and family.
49. Continuity is especially vital where young families are involved. It cannot be emphasised enough how young children require the **stability / confidence / trust** which would result from continuity of care of a young parent. The same continuity of care must also assist the home care staff to build up that **trust / confidence with all members of families concerned**.
50. 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 exhibit disturbed behaviour nurses trained in mental health nursing or learning disability nursing should be involved in their care or should be available to provide advice and support for the caring team.

### ***Palliative care***

- 51.** The decision about the best place for patients to receive palliative 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.
- 52.** Palliative care should be provided as near as possible to patients' homes. Patients should be cared for in an environment appropriate to their age, especially younger patients. The key worker should identify suitable places and help the family to choose the most suitable for their own circumstances. Advice regarding suitable placement according to current and anticipated needs can be sought from the local Regional Expert and also the National CJD Co-ordinator.
- 53.** 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***

- 54.** 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.

55. Families caring for patients at home should be offered advice on the practical care of patients, including movement and handling, and the management of challenging symptoms.
56. The rapidity of progression of CJD means that any necessary aids or equipment, including incontinence pads, should be provided rapidly when they become needed. The regular assessment of care should both identify proactively and plan to meet these needs in advance to avoid undue delay causing distress to the patient or their carers.
57. Many families would benefit from the provision of respite care and night nursing services. These should be provided promptly when needed. The regular assessment of care should plan for these services which will need to be arranged on an individual basis.
58. As carers, families have a right to have their own needs assessed and any necessary help provided.

### ***Other support for families***

59. 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.
60. Many families suffer financially when caring for a patient with CJD. They should receive information on benefits. In particular, patients eligible for Attendance Allowance (over 65) or Daily Living Allowance (under 65) may qualify for urgent payment under special rules.

61. All families need a high level of emotional support, during and after the patient's illness. Professionals providing care of patients should provide informal support, continuing after the patient dies. General Practitioners can have a particular role in this continued support of the family.
  
62. 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***

63. Professionals caring for patients are also often distressed by the experience. They should be offered support and counselling if required. There is a perception by some that there is a stigma attached to admitting to the effects of stress or trauma. Experience shows that staff accept support more readily from sources outside their own employer's management arrangements. South and East Belfast Trust have developed a Staff Care service which offers a confidential, professional counselling service to a wide range of employers, including many Trusts. All Trusts should be encouraged to identify similar support services which can be accessed by those staff requiring confidential assistance.

### ***Maintaining confidentiality***

64. Professionals involved in the care of patients should be aware of the media interest in CJD. It is important to maintain patient confidentiality. For example, a code number rather than a name should be used on clinical samples. No information should be given about patients over the telephone until the caller's identity has been verified.

### ***Handling the media***

65. Some families of patients with CJD have been subject to unwanted media attention. Trusts should have a media strategy in place and should identify a person(s) who would have responsibility to protect the patient/patients' family from unwarranted attention. Trust professionals should be aware of and follow this strategy in any contact with the media in order to provide sanctuary to patients/patients' families from intrusive media behaviour and often ill informed and stigmatising media coverage. In the specific case of vCJD, the Human BSE Foundation can offer advice to families on dealing with the media.

### ***Funeral arrangements***

66. 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 95-101 for advice on infection control for undertakers).



### III. INFORMATION NEEDS AND COMMUNICATION

67. 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.
68. 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. It would be good practice to keep a multi-disciplinary record which accompanies the patient regardless of setting and in which relevant health and social care information is kept.

#### *Communicating the diagnosis*

69. 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.
70. Patients' GPs should be kept informed of the patient's condition and care arrangements. GPs have a role in palliative care, and in continued support of the family.

### ***Likely disease progression***

71. 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.
  
72. 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***

73. The possibility of post mortem, including the potential involvement of the coroner and the process of an inquest, 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 which 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 and retention of tissues after death. Relatives should be informed that post mortem is not compulsory, unless the coroner is involved and deems it necessary, but is needed to confirm the diagnosis and help in research into this disease.



#### IV. SAFE WORKING AND PREVENTION OF INFECTION

74. Detailed guidance on infection control of CJD in a healthcare setting is included in the ACDP/SEAC document ‘Transmissible spongiform encephalopathy agents: safe working and the prevention of infection’ A DHSS Circular ‘Variant Creutzfeldt Jakob Disease (vCJD): 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).
75. The guidance identifies three patient risk groups. The following points are summarised from the guidance on care of patients with known or suspected CJD. (ACDP/SEAC Guidance).
76. 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 ACDP/SEAC Guidance.
77. **Current evidence suggests that normal social or routine clinical contact does not present a risk to healthcare workers, families or others.**
78. Special precautions are only required for handling high risk tissues. These are, brain, spinal cord including cerebro-spinal fluid, or eye tissue and for vCJD, lymphoreticular tissue.
79. 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.

### ***Infection control precautions in hospital***

- 80.** 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.
- 81.** There is no evidence of infectivity in saliva, urine, vomit or faeces. 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.
- 82.** In patients with known or suspected CJD, should sheets become contaminated with cerebrospinal fluid or other high-risk material they should be disposed of by incineration.

### ***Clinical procedures***

- 83.** There is uncertainty about the risk of transmission from blood, but normal infection control procedures 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.
- 84.** Should any accident involving sharps or contamination of abrasions with blood or body fluid occur, the wound 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.

- 85.** Blood spillages should be treated in the normal way. 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 dispose of by the normal route. For the safe handling of infectious material double bagging is advisable. Then clean the area with detergent and warm water and dry.
- 86.** For all other body fluid spillage eg. Urine/vomit/faeces, absorb fluid with paper towels. Dispose of waste into yellow clinical waste bags and dispose of by the normal route. Clean the area with detergent and warm water and dry. The application of a disinfectant solution is unnecessary.
- 87.** The procedure described in paragraph 85 will not inactivate prions. For spillages of high risk materials (paragraph 78), cover with 20,000 ppm available chlorine of sodium hypochlorite for one hour or 2M sodium hydroxide for one hour (see Annex B in ACDP/SEAC Guidance). Where fomites are contaminated with high risk materials these must be disposed of by incineration. For the safe handling of high risk material double bagging is advised.
- 88.** Routine clinical specimens should be stored and transported as local policy dictates. It is important to ensure that all specimens are clearly coded and securely bagged for transportation.
- 89.** 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.

### ***Infection control precautions in the home setting***

90. 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.
91. Families should not be dissuaded from ordinary contact with patients but should wear gloves and aprons if handling body fluids e.g. urine, vomit, faeces or blood.
92. 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.
93. Families should be provided with plastic bags for any clinical waste Materials and sharps containers if appropriate.
94. 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 from the home are put in place.

### ***After death***

- 95.** After death, the body should be placed in a body bag and removed to the mortuary using normal infection control measures.
- 96.** If a post mortem examination 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. In this circumstance, contact with the body should be minimised following post mortem.
- 97.** The 1998 guidance “Transmissible Spongiform Encephalopathy Agents: Safe Working and the Prevention of Infection” should be consulted for advice on precautions to be applied during post mortem examination.
- 98.** 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.
- 99.** Embalming is contraindicated.
- 100.** Relatives need not be discouraged from viewing the body or from superficial contact such as touching the face.
- 101.** 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

### **LOCAL REGIONAL EXPERT**

Dr Tom Esmonde  
Consultant Neurologist  
Department of Neurology  
Royal Victoria Hospital  
Grosvenor Road  
Belfast BT12 6BA  
Tel: **028 9089 4921**

### **CJD SUPPORT NETWORK**

The CJD Support Network is a voluntary organisation set up to provide help and support for patients with all types of CJD and their families. 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. This initiative extends to Northern Ireland.

Gillian Turner  
National CJD Coordinator  
CJD Support Network  
Birchwood  
Heath Top  
Ashley Heath  
Market Drayton  
Shropshire TF9 4QR  
Tel: **01630 673 993**

## **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  
299 Bonnington Drive  
Deaglesham  
Glasgow G76 0NH  
**Tel (Helpline): 0191 3894157**

## **NATIONAL CJD SURVEILLANCE UNIT**

The National CJD Surveillance Unit 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 Unit does not undertake the management of patients but is able to offer an opinion on whether a patient has CJD, and give advice on investigations.

Professor R G Will  
National CJD Surveillance Unit  
Western General Hospital  
Crewe Rd  
Edinburgh EH4 2XUT  
**Tel: 0131 332 2117**  
**Fax: 0131 343 1404**

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.

### **THE PRION UNIT**

The Prion Unit at St Mary's Hospital, London is a tertiary referral and research centre for CJD.

Kathryn Prout  
Clinical Nurse Specialist  
Prion Unit  
Department of Neurology  
St Mary's Hospital  
Praed St  
London W2 1NY  
Tel: **0171 886 6883**



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## ANNEX A

### **ISSUES TO BE CONSIDERED AT CASE CONFERENCE**

1. Assessment and reassessment of patients' care needs involving:
  - nursing care
  - social services input
  - occupational therapy
  - dietetics
  - physiotherapy
  - speech and language therapy
  - laundry facilities
  - specialist incontinence services
  - palliative care
  - voluntary sector services
2. Assessment and reassessment of family/carers needs for practical help and childcare.
3. Co-ordination of professionals providing care including voluntary sector services.
4. Provision of resources to meet identified needs.
5. Advice on financial help/benefits.
6. Flexibility of care package to include availability of respite care and 24 hour community care for both patient and family.
7. Provision of information and emotional support to patient/family/carer.
8. Assistance with end of life decisions and help with funeral arrangements.
9. Emotional support/assessment for formal counselling for families/carers after the patient dies.

**ANNEX B****MEMBERSHIP OF WORKING GROUP**

**Dr Philip McClements,**  
Deputy Chief Medical Officer, DHSSPS - Chairperson

**Mrs Mary Hinds,**  
Nursing Officer, DHSSPS

**Mrs Liz Atkinson,**  
Nursing Director, Northern Ireland Hospice

**Mrs Maeve Girvan,**  
Patient and Carer Representative

**Dr Anne Montgomery,**  
Consultant Psychiatrist, Mater Infirmorum Hospital

**Mr John Park,**  
Social Services Inspector, DHSSPS

**Dr Liz Mitchell,**  
Principal Medical Officer, DHSSPS

**Dr Tim Wyatt,**  
Consultant Microbiologist, DHSSPS

**Dr Ian McMaster,**  
Medical Officer, DHSSPS