6. Diagnosis, treatment and care of vCJD patients

Introduction

6.1 The main aim of the CJDSU was to identify any change in the clinical, pathological and epidemiological characteristics of CJD that might be linked to the occurrence of BSE. The first part of this chapter details how, between 1995 and 1996, the CJDSU collected evidence of the emergence of a new variant of CJD (vCJD), the signs of which included an earlier age of onset than classical CJD, early psychiatric symptoms, a long duration of illness, and an absence of the characteristic EEG pattern seen in classical CJD. It goes on to describe the methods used to diagnose the disease along with recent advances in this area. The second part of the chapter describes the treatment and care of vCJD patients and addresses some of the issues that were and are still being faced by vCJD victims and their families.

6.2 Much of the evidence referred to in this chapter does not distinguish between events before and after 20 March 1996. It would not be practicable to attempt this exercise; nor do we consider that our Terms of Reference require us to do so. Problems experienced and solutions found after 20 March 1996 throw light on the experience before that date and are relevant when considering the lessons to be learned from events falling within the period with which we are concerned.

Symptoms of vCJD

6.3 On 21 March 1996, the CJDSU circulated to all neurologists in the UK a description of the clinical and pathological features of the new phenotype. The clinical features of the new phenotype of CJD described in the circular were as follows:

i. An early age of onset or death (average 27.6 years, range 18–41 years).

ii. A prolonged duration of illness (average 13.1 months, range 7.5–24 months).

iii. A predominantly psychiatric presentation including anxiety, depression, withdrawal and progressive behavioural changes.

• The first evidence of neurological involvement in four patients was dysaesthesiae in the limbs and/or face.

• Development of a cerebellar syndrome with problems with gait and limb muscle coordination after a period of weeks or months.

390 S61 Will para. 3
391 EEG – electroencephalogram (a measurement and recording of electrical activity in the brain)
392 IBD3 tab 9
393 Dysaesthesiae – unpleasant abnormal sensations
Development of forgetfulness and memory disturbance, often late in the clinical course, which progressed to severe cognitive impairment and a state of akinetic mutism\(^{394}\) in the majority of cases.

Development of muscle twitching or spasms in the majority of patients (myoclonus), preceded by purposeless involuntary movements in some (chorea), with EEG appearances typical of sporadic CJD absent.

6.4 The circular noted that there was likely to be an increase in psychiatric referrals as the early symptoms of vCJD were ‘relatively non-specific’. It also mentioned that early identification might depend on a history of evolving psychiatric disturbance followed by clear evidence of a cerebellar syndrome and/or memory disturbance.\(^{395}\)

**Diagnosis of vCJD by the medical profession**

6.5 In his written evidence to the Inquiry, Dr Will observed that the clinical diagnosis of CJD (as with many other neurological disorders) was not dependent on the application of standardised research diagnostic criteria but was a matter of clinical training and experience.\(^{396}\) Thus, the identification of patients with vCJD was largely dependent on the diagnostic expertise of the medical professional.

**General practitioners**

6.6 In most cases, following the manifestation of vCJD symptoms, the patient’s first contact with the medical profession was with the family general practitioner (GP). In evidence to the Inquiry, the families of vCJD victims described some of the symptoms they observed that influenced their decision to consult a GP. These included depression,\(^{397}\) coordination problems,\(^{398}\) memory loss and mood swings,\(^{399}\) pins and needles and pains in the limbs,\(^{400}\) bad headaches,\(^{401}\) cold extremities,\(^{402}\) pain in the feet,\(^{403}\) rashes\(^{404}\) and short-term memory loss.\(^{405}\) In all cases, the symptoms worsened over a relatively short space of time and the GP referred the patient on to a psychiatrist or neurologist. In all cases, the patients were ultimately referred to a neurologist.

**Psychiatrists**

6.7 The part played by the psychiatrists in the diagnosis and management of cases of vCJD is summarised in a large study by the CJDSU of the psychiatric features of 33 cases in the *Psychiatric Bulletin*\(^{406}\). The article noted that the majority of vCJD cases had seen a psychiatrist, often as the first specialist referral, but that the non-

\(^{394}\) Akinetic mutism – paralysis and inability to speak
\(^{395}\) IBD3 tab 9
\(^{396}\) S61 Will para. 16
\(^{397}\) T72 pp. 65, 67, 71 and 78–9
\(^{398}\) T73 pp. 1–2
\(^{399}\) T72 p. 73
\(^{400}\) T72 p. 73
\(^{401}\) T72 pp. 75–6
\(^{402}\) T72 pp. 76, 79 and 132
\(^{403}\) T72 pp. 63 and 73
\(^{404}\) T72 p. 77
\(^{405}\) T72 p. 82
specific characteristics of the psychiatric symptoms of vCJD made early recognition difficult or impossible.

6.8 The cases took an average of 8 months to be referred to a psychiatrist – where this was the first specialist referral – after onset of illness (ranging from 2 weeks to 16 months). Seventeen of the cases were initially referred to a psychiatrist, and eight cases involved psychiatrists after referral to a neurologist. The average time between consultation with a psychiatrist and referral to a neurologist was 9 weeks (ranging from one day to 32 weeks). Similarly, where the initial referral was to a neurologist, the average time before referral to a psychiatrist was 10 weeks (ranging from 2 to 38 weeks).

6.9 All but one of the 33 cases exhibited psychiatric symptoms at an early stage and for most cases these symptoms were the first manifestation of the disease. The main symptoms were depression, anxiety, withdrawal, aggression, hallucinations and delusions. In addition, loss of memory was an early symptom in four cases. Two patients had suicidal thoughts. Neurological symptoms were often also present during the early ‘psychiatric’ phase. Persistent and severe sensory complaints were noted as a symptom in 17 cases, including pain and abnormal sensations in hands, feet, face, mouth and lumbar region. These are discussed more fully in the section on neurologists below.

6.10 Despite these neurological symptoms, many of the patients were initially thought to be suffering from a psychiatric, as opposed to neurological, disorder. The predominantly psychiatric symptomatology meant some patients were referred for psychological testing even after being seen by a neurologist. In most cases the diagnosis given was depression, and 19 cases were prescribed antidepressants, often initially by their GP. Organic disease was suspected in a minority of cases with early cognitive impairment, and with those displaying forgetfulness. In a small number of cases a psychotic illness was suspected. Neuropsychological assessments carried out in ten cases recorded clear evidence of cognitive impairment, but the majority of these examinations were performed after the development of earlier neurological signs. Two cases had already demonstrated cognitive impairment, with one case showing forgetfulness as an initial symptom.

6.11 The Psychiatric Bulletin article noted that tests carried out during the psychiatric phase were invariably normal and thus ‘almost always unhelpful’ because they did not point to a neurological disease. These tests included EEG and magnetic resonance imaging (MRI). The tests that had been performed before neurological referral detected no abnormalities. (These tests are discussed more fully at paragraphs 6.22–6.39.)

6.12 In conclusion, the article noted that the diagnosis of vCJD might not be possible in the early stages of the illness, because of the difficulty of distinguishing between psychiatrically determined neurological features and those symptoms due to an underlying organic disease. This was compounded by the fact that some

407 Ibid.
408 Ibid.
409 Organic disease – a disease process which occurs as a result of a demonstrable anatomic or physiologic abnormality, eg, brain tumour, Alzheimer’s disease, etc
410 Cognitive impairment – impairment of the mental processes involved in knowing, thinking, learning and judging
411 Psychotic illness – where there is an impairment of perception of reality
413 Ibid.
patients showed transient improvement following treatment with antidepressants. The diagnosis of vCJD was often considered not possible until the occurrence of overt neurological signs. However, the article identified some symptoms that might aid the diagnosis of vCJD during the ‘psychiatric’ phase:

The identification of cognitive impairment in addition to the psychiatric symptoms may raise the possibility of an underlying organic disorder in some cases. Persistent painful limb or joint pain, dysaesthesia\(^4\) or paraesthesia\(^5\) in combination with the psychiatric symptoms is unusual and may raise the suspicion of new variant CJD. Such symptoms include painful sensory disturbance in the hands and feet, in the face or mouth, or in discrete areas of the trunk. Some patients describe dysaesthesia in a similar distribution and some limb or joint pain. The persistence of these symptoms and their severity is distinct from the transient paraesthesia that may accompany anxiety and hyperventilation. Other suggestive neurological symptoms during the psychiatric phase include gait imbalance, visual disturbance, dysarthria\(^6\) dysgeusia\(^7\) and involuntary movements, although it is understandable that these symptoms have in some cases been attributed to side effects of medication.\(^8\)

6.13 The article noted that any delay in diagnosis was distressing to the relatives of patients but that the evidence suggested that the psychiatric community was referring cases of vCJD promptly to neurologists.\(^9\)

Neurologists

6.14 In 1997, the CJDSU published an article describing the neurological features of the first 14 cases of vCJD and the tests used to provide a diagnosis.\(^10\) The description of the diagnostic tests is covered at paragraphs 6.22–6.39. Here, the clinical neurological features of vCJD of the first 14 cases are explored.

6.15 The article noted that although a minority of cases suffered from mild unsteadiness of gait from an early stage, clear neurological symptoms were not apparent before an average of 6½ months after onset of illness, ranging from 4 to 24½ months. The first neurological symptom was generally ataxia,\(^11\) including cerebellar, limb or gait ataxia. The ataxias occurred in isolation and in combination with involuntary movements, pyramidal signs,\(^12\) limited reflexes or sensory signs. The remaining cases first developed either pyramidal signs (with or without problems with language) or involuntary movements, with unsteadiness of gait apparent within weeks.\(^13\)

\(^4\) An unpleasant, abnormal sensation
\(^5\) Unusual feelings, apart from increase or loss of sensation experienced by a patient without any cause, such as hot flushes, numbness, tingling or itching
\(^6\) Problems with speech
\(^7\) Problems with taste
\(^9\) Ibid.
\(^11\) Ataxia – failure of muscle coordination; unsteady gait
\(^12\) Pyramidal signs – increased muscle tone and abnormal reflexes
6.16 Usually, the symptoms progressed quite rapidly to include global cognitive impairment, involuntary movement, progressive immobility, unresponsiveness, mutism and urinary incontinence. In the days before death, the patients were akinetic mutes and three developed cortical blindness. On average, there was 6 months’ lapse between developing unsteadiness and becoming bedridden (range 2½–12½ months). On average there was 1½ months between becoming bedridden and death (ranging from 1 week to 6 months). Two patients remained alive 6½ and 18 months respectively after becoming bedridden.424

6.17 For those with a long delay in the onset of neurological symptoms, these initial symptoms were prolonged with personality change or forgetfulness followed by sensory disturbance.425 Most cases developed primitive reflexes, cerebellar and pyramidal signs: all had persistent involuntary movements. Seven cases were noted to have upgaze paralysis, a condition which prevents a patient from looking upwards and which is an uncommon feature of CJD.

6.18 In oral evidence, Professor Martin Rossor of the National Hospital of Neurology and Neurosurgery, London, described the difficulties faced by clinicians when diagnosing vCJD.426 Professor Rossor had established an inpatient and outpatient service specialising in early onset and unusual dementias. He also led the Dementia Research Group and held an MRC Special Project Grant specifically to study vCJD.

6.19 Professor Rossor commented upon the problems encountered in the diagnosis of CJD in young patients. He began by defining what is meant by ‘early onset dementia’:

Classically, early onset is below the age of 65, which has more to do with the common retirement age than any biological distinction. In general we would see people with dementia starting sometimes as young as their 20s, through to people in their 50s. We do, of course, see older people as well but that is the special area of interest.427

6.20 Professor Rossor then commented upon the problems of obtaining a clear diagnosis in cases of early onset dementia:

. . . We always try to give a name. And often, because we have to work with the tools that we have, one always tends to try to fit something into a category. In this area it is very difficult. I think two points. One is that I would think that approximately, and this is my first thought, perhaps 30 per cent of cases I see I may not be able to arrive at a clinical diagnosis. So one leaves it in very broad terms. Even when one can examine the brain by cerebral biopsy, which we do sometimes have to do, or can examine the brain after death at autopsy, one is sometimes still left without being able to make a diagnosis.

. . . If we take the group below the age of 65 taking the orthodox view of early onset. Approximately 50 – if you just allow me to think through this. Probably about 40 per cent are Alzheimer’s disease . . .

424 Ibid.
425 Ibid.
426 T72
427 T72 p. 12
Another large group are those we loosely refer to as the frontotemporal dementias. Those are characterised by the fact they have early behavioural change followed often by changes with language. That can create considerable diagnostic difficulties. That group consists of four different diseases that we tend to recognise, and they include a disorder called frontal lobe degeneration of the non-Alzheimer type. The second is Pick’s disease. The third is referred to as the frontotemporal degeneration and motoneurone disease complex, and another type that we increasingly identify is that of frontotemporal degeneration with what are referred to as ubiquitin inclusions. There are other diseases that are very rare. Our ability to make a specific diagnosis is very limited unless we carry out a brain biopsy. That probably accounts for about 80 per cent of cases of early onset, although these figures are very broad. The remainder are a variety of disorders which will include CJD, will include vascular disease where there are multiple strokes due to a variety of causes, alcoholism and some other rare disorders.428

6.21 Professor Rossor also referred to problems in the early diagnosis of CJD in the young due to the initial presentation of cognitive impairment:

Cognitive impairment or dementia which just refers to a generality of cognitive impairment, not just memory lack, visuoperceptual problems, thinking, planning, is of course seen with many, many different causes and the list runs into hundreds of potential diseases. And many are relatively easily excluded right at the beginning, but the path to a differential diagnosis of an early onset unusual dementia can be a very long one.429

Tests performed

6.22 Most of the tests performed on vCJD victims while they were alive occurred when the patient was in the care of a neurologist. Generally, the tests only proved useful in excluding other possible conditions, and even now a conclusive diagnosis of vCJD is not usually possible until post-mortem neuropathological examination of the brain has been performed.

6.23 In the 1999 *Psychiatric Bulletin* article on the psychiatric features of the first 14 vCJD patients, it was observed that ‘limited evidence suggests that investigations such as EEG or brain imaging are unlikely to provide useful diagnostic information during the “psychiatric” phase of these diseases’.430 The families of the victims of vCJD often mentioned the distress caused by the failure of the medical profession to diagnose vCJD until late in the illness. The recent development of a tonsil biopsy test (see paragraphs 6.37–6.39 for further details) for protease-resistant prion protein may allow earlier diagnosis. Here, a range of tests, reported in the CJDSU articles and by expert witnesses, is discussed.

428 T72 pp. 13–4
429 T72 p. 18
Blood tests

6.24 Blood tests are generally carried out on suspect CJD patients to detect or exclude haematological or biochemical disturbances, and in particular to detect abnormalities of kidney and liver function, and calcium metabolism, all of which are associated with cognitive impairment. The level of vitamin B12 and folate are measured, as low levels of vitamin B12 sometimes manifest themselves as mental changes. Thyroid function is also tested to exclude myxoedema (underfunctioning of the thyroid) and Hashimoto’s disease (an autoimmune disease in which the whole of the thyroid gland is diffusely enlarged and firm).

6.25 An examination is made for antibodies against the organism causing syphilis and the presence of systemic lupus erythematosus. If there is a possibility, based on clinical history, of exposure to heavy metal such as lead, mercury or manganese, tests are carried out to exclude heavy metal poisoning.

Genetic analysis

6.26 Before a diagnosis of vCJD can be made, the diagnosis of familial CJD must be ruled out. Familial CJD is associated with mutations of the prion protein gene. DNA is therefore extracted from the blood of all suspect cases and screened for prion protein gene mutations. No mutations have been identified in cases of vCJD. In all cases the genotype is determined at codon 129 of the prion protein gene.

Lumbar puncture

6.27 Lumbar puncture is a procedure for removing cerebrospinal fluid (CSF) from the lumbar region of the spinal canal and is used in the diagnoses of some diseases of the nervous system. The fluid is tested for evidence of inflammation and syphilis. It can be used to exclude the diagnosis of multiple sclerosis.

6.28 It has been shown in the USA that a specific protein marker (14-3-3 protein) can be detected in the CSF from patients with CJD, raising the possibility that it can be used as a specific marker for disease. The 1997 *Lancet* article noted that out of five vCJD patients analysed for the presence of protein 14-3-3, two were positive and three negative. However, the test cannot differentiate between the different types of CJD.
Computerised axial tomography (CT) scan

6.29 This technique produces cross-sectional images of the body and allows examination of the brain to assist in neurological diagnosis.\textsuperscript{442} The results of this test for 10 of the first 14 cases of vCJD were discussed in the 1997 \textit{Lancet} article.\textsuperscript{443} Eight patients had normal scans. The two abnormal scans had non-specific abnormalities: calcium deposits in one part of the brain and slightly enlarged areas of the brain.

Electroencephalogram (EEG)

6.30 An EEG records electrical activity in the brain. A number of small electrodes are placed on the scalp and the pattern of activity is recorded.\textsuperscript{444} The 1997 \textit{Lancet} article noted that the characteristic EEG pattern of sporadic CJD was not seen in the vCJD cases.\textsuperscript{445} Several presented with normal patterns even though they had cognitive impairment, cerebellar signs and involuntary movements. Of those which had abnormal EEGs, all showed slow-wave activity which deteriorated as the illness progressed:

The majority of cases thought to have a functional psychiatric illness developed an abnormal EEG within 3 months of their psychiatric diagnosis. However, one patient had a normal recording 7\frac{1}{2} months after the diagnosis of schizophreniform psychosis.\textsuperscript{446}

Magnetic resonance imaging (MRI)

6.31 MRI is another three-dimensional way of looking at the brain. The head of the patient is placed in a magnetic field which causes certain atomic nuclei to align in the direction of the field. Pulses of radio-frequency radiation are applied and interpretation of the radiation absorbed and re-emitted allows an image of the brain to be built up.

6.32 Traditionally, the role of MRI in patients with suspected CJD has been to exclude other conditions. In his statement to the Inquiry, Professor Rossor observed that the scan is used to ‘exclude structural lesions such as tremors and blockage of the ventricular system, inflammatory disorders and may identify areas of tissue loss or atrophy.’\textsuperscript{447}

6.33 Recent work, however, has suggested that MRI can be used in the differential diagnosis of suspected vCJD. Analysis of MRI scans of confirmed vCJD patients revealed high signal in the posterior thalamus (pulvinar).\textsuperscript{448} The signal was found to be specific, being present in 78 per cent of the 36 vCJD patients examined but in none of the 57 control patients. The presence of this specific signal on MRI scans

\textsuperscript{442} M26A tab 4, p. 8
\textsuperscript{444} M26A tab 4, p. 8
\textsuperscript{446} Ibid
\textsuperscript{447} S194 Rossor para. 4
may in the future facilitate the diagnosis of this form of CJD during the clinical course, avoiding the need for invasive diagnostic procedures.

**Biopsies**

**Brain biopsy**

6.34 One test that can be used to diagnose vCJD before death is the histopathological examination of a brain biopsy.449 This examination aims to detect the characteristics of vCJD: ‘florid plaques’, abundant prion protein deposition, sponginess and severe glial cell450 proliferation in the thalamus.451

6.35 However, the ante-mortem examinations of brain biopsies are not thought to be reliable. Dr Will described the main limitation of the brain biopsy. Only a very small piece of brain tissue is removed and this sample might be ‘from an area of the brain that is not affected fully by the pathological process, and you may sometimes sample an area that is normal, which may subsequently become abnormal. So a negative test does not exclude a diagnosis’.452

6.36 In his oral evidence to the Inquiry, Professor Rossor commented upon the use of brain biopsies in the diagnosis of rare dementing illnesses and emphasised that the procedure carried risk and was not performed if other tests revealed a probable diagnosis.453

**Tonsil biopsy**

6.37 Histopathological examination of tonsil biopsies has been suggested as a useful diagnostic test for those with suspect vCJD. An experimental study to examine the reliability of this test was published in 1999.454 Tonsil biopsies were performed on 20 suspect CJD cases. The biopsies were then examined histopathologically for the characteristic protease-resistant prion protein and prion protein glycosylation pattern seen in vCJD.

6.38 Eight tonsil biopsies tested positive for the protease-resistant prion protein; all these patients were subsequently confirmed as vCJD cases, or their subsequent course of disease was highly consistent with vCJD. All tonsil biopsies negative for the protease-resistant prion protein were later shown to be from cases of either sporadic CJD or iatrogenic CJD. There were no false negative results in vCJD cases and each showed the glycosylate pattern characteristic of vCJD.

6.39 The study concluded that the protease-resistant prion protein could be identified in all vCJD cases tested so far by tonsil biopsy. In practice, the tonsil biopsy might obviate the need for a brain biopsy sample to be taken. However, patient acceptability of tonsil biopsy is low.
Clinical history – personal and family

6.40 When the referring neurologist notifies a suspect case of CJD to the CJDSU, the Unit generally carries out a detailed personal and family clinical history based on a questionnaire. This is described earlier in this volume.

6.41 Professor Rossor pointed out that a detailed family history is obtained in all cases to ascertain if the condition is genetic. If this is suspected, tests are then made for genetic mutations known to be associated with familial dementia such as Huntington’s disease, familial Alzheimer’s disease, frontotemporal dementia and the familial prion diseases which include disorders such as familial CJD, Gerstmann-Sträussler Syndrome (GSS) and Fatal Familial Insomnia (FFI). However, not all cases of inherited CJD have a positive family history. The patient could be the first in a family to inherit the mutation through parental germ cells.

Treatment and care of vCJD patients

6.42 To date, over 70 people have tragically died from the horrific disease that is vCJD. The families of the victims of vCJD have demonstrated great courage in assisting the Inquiry by recounting the terrible and painful events leading up to and following the death of their loved ones. The families have tirelessly pursued the government for answers to the question of how vCJD arose and were instrumental in campaigning for the BSE Inquiry.

6.43 This part of the chapter addresses some of the issues that were are still are being faced by the victims of vCJD and their families, both during illness and following death. The Inquiry heard oral evidence from 15 family members on these and other issues, and many other family members contributed through written statements. We have also received material from those providing care and from lawyers representing the victims’ families.

6.44 The information provided to the Inquiry by the families has been invaluable in giving a ‘snapshot’ of the effects of the disease on the victim, on the family and on medical staff and carers of vCJD patients.

6.45 The information has not been gathered scientifically and is not intended as a clinical study of care and treatment. The evidence provided by the families is necessarily subjective, as are the responses from hospitals and doctors. The doctors and hospitals were understandably loathe to compromise patient confidentiality and many chose not to respond to the evidence provided to the Inquiry by the families. It is often the case that things that went wrong in the course of care and treatment stay in the mind far more than things that went right. Each case is different and each family had a different experience, just as the disease progressed slightly differently from person to person.

6.46 It has been possible, however, to identify a number of similarities in the experiences of the vCJD victims and their families. This section, therefore, aims to give an overview of what the care and treatment of vCJD patients involved and how it changed as the disease progressed.

455 S194 Rossor para. 3
6.47 We examine care and treatment that was good as well as the all too frequent scenario of care and treatment that was less than adequate and problems encountered by the families.

Progression of the disease and stages of care

6.48 The first part of this chapter sets out the clinical symptoms of vCJD and discusses the means by which the diagnosis of vCJD by the medical profession has been made. Just as various stages of the disease have been identified, so too have there been stages in the treatment and care of vCJD patients which arise according to the progression of the disease.

6.49 The stages of the disease and the consequent stages of treatment and care are summarised below. This summary is not exhaustive and of course not every case has followed the same pattern. However, this summary reflects the experience of many of the families who have submitted evidence to the Inquiry:

i. Examination and treatment by the patient’s General Practitioner (GP) following the onset of non-specific symptoms.

ii. Referral from the GP to a psychiatrist and/or a neurologist.

iii. Admission to hospital for testing by the psychiatrist and/or neurologist.

iv. Notification of the national Creutzfeldt-Jakob Surveillance Unit (CJDSU) and a preliminary diagnosis of vCJD made.

v. Treatment and care of the patient following diagnosis of probable vCJD.

vi. Post-mortem and burial/cremation issues.

vii. Death certification.

viii. Consideration of whether an inquest, or fatal accident inquiry in Scotland, will be held.

Initial care and treatment by the General Practitioner (GP)

6.50 GPs were normally the first point of contact for people suffering from the first symptoms of vCJD. The inevitable difficulties surrounding the diagnosis of a very rare disease were evident at this stage, according to the experiences recounted by most of the witnesses. As mentioned earlier in this chapter, the symptoms of the disease in the early stages were relatively unremarkable.

6.51 The mother of a patient described in her witness statement how her daughter ‘became withdrawn, forgetful, constantly tired and lost a lot of weight. She also constantly complained about pains in her legs’. Similar symptoms were experienced by many of the vCJD patients.

6.52 Commonly GPs arranged for tests to be undertaken, and generally the results failed to reveal any abnormality or disorder.
6.53 The difficulty in settling upon a diagnosis is clearly depicted in the witness statement of a GP who wrote:

The history of the presenting complaints was difficult to attribute to a specific diagnosis . . . [The patient] could be suffering from depression but I could not attribute all of her symptoms to this diagnosis. Her neurological symptoms were not precise and did not clearly fall into any recognised neurological disorder at that time. I noted the results of her tests and the equivocal plantar response and did not feel that I was able to make a firm diagnosis . . . I concurred that nervous exhaustion was the most appropriate diagnosis that we could make at the time.\textsuperscript{457}

6.54 This inability to pin down the symptoms to a recognised medical complaint was a common story for GPs.

6.55 A GP who gave oral evidence told us that in the case that he dealt with, the patient initially consulted him complaining of ‘increasing anxiety and some lowering of mood’ in addition to ‘a bad taste in her mouth . . . and she felt that was affecting her appetite’.\textsuperscript{458} Counselling was organised initially. Over the next few months evidence of depression was identified and an antidepressant prescribed.\textsuperscript{459} The patient’s condition regressed and eventually the GP decided that he needed specialist assistance. He said:

I did not feel I could manage [the patient’s] case in general practice any more. I actually said that I did not feel I had ever seen a case like [the patient’s] before. I still was confident that her diagnosis was psychiatric. I was not entertaining anything else, but I felt I needed help of more specialist services.\textsuperscript{460}

6.56 The account given in the above paragraph seemed to be a widespread experience for GPs. It was only when the symptoms progressed from the commonplace to the more extreme that GPs either referred their patients to a specialist, typically a psychiatrist or a neurologist, or families took matters into their own hands and sought specialist treatment direct.

Referral of patient to a psychiatrist or a neurologist

Psychiatrists

6.57 Psychiatrists to whom patients had been referred reported similar difficulties in diagnosis to the GPs. The experience of a number of psychiatrists is epitomised in one particular case where the patient initially met the criteria for a depressive disorder and was prescribed antidepressant drugs. Despite this, efforts were made to find a physical cause for the condition. Subtle neurological signs were seen but were attributed to the medication that the patient was taking. The patient’s condition deteriorated rapidly and she was referred for neurological assessment.\textsuperscript{461}

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\textsuperscript{457} S598 Harrison response p. 2  \\
\textsuperscript{458} T2 p. 72  \\
\textsuperscript{459} T2 p. 74  \\
\textsuperscript{460} T2 p. 77  \\
\textsuperscript{461} S236 paras 21–4
\end{flushleft}
Neurologists

6.58 Often by the time a patient was referred for neurological examination, a psychiatric examination had been conducted already, without success, and organic symptoms had been identified, thus prompting neurological examination.

6.59 It was commonly during the course of neurological examination and testing that the possibility of vCJD was raised. At this point the CJDSU was usually contacted and a preliminary diagnosis of vCJD made.

Admission to hospital for testing by the psychiatrist and/or neurologist

6.60 As described above, one of the biggest problems when the first cases of vCJD arose was that of diagnosis. Because the condition had not been seen in young patients before, GPs had difficulty identifying what kind of disorder patients were experiencing and thus who to refer them to. Because some of the earliest signs and symptoms were psychiatric in nature, several patients were admitted to psychiatric wards or specialist psychiatric hospitals. Treatment on these wards, including the nursing care, appears at times to have inappropriate.

6.61 It is noted that at this stage vCJD was not usually considered to be a possibility – medical and nursing staff were treating what were considered to be psychiatric symptoms. This problem was highlighted in the report ‘Patients with new variant Creutzfeldt-Jakob disease and their families: care and information needs’:

The organisation and type of care provided in psychiatric units were often inappropriate for patients with deteriorating physical problems. Similarly, acute medical and neurology units found it hard to manage patients with severe psychiatric disturbance.462

6.62 A number of the hospitals have described how they struggled with the care of the patients admitted to their care.463

6.63 One particular hospital described the manner in which the hospital attempted to deal with the problem. In this case the hospital readily admitted that nursing the particular patient was a ‘challenge’ for their nursing staff. The patient would wander off if not constantly supervised. The nursing staff became frustrated by the patient’s constant wailing day and night, which is now known as a symptom of the disease, but for the nursing staff at the time, seemed to have no apparent cause. A support group was subsequently set up by the hospital for the staff so that a strategy could be established to identify the patient’s needs. Advice was sought from a Mental Health Consultant to establish how to care for the patient. As a result, additional nursing support was able to be provided for the ward.464

6.64 A number of complaints made about hospitals were able to be explained by the hospitals when statements were requested from them in response to the families’ statements. Similarly, a number of the hospitals remarked that they did not realise that the families had had concerns about the hospital care until they read the

462 M26A tab 10
463 S567 Blundell response para. 10; S580 Robinson response
464 S580 Blundell response para. 9
families’ statements.\(^\text{465}\) Therefore, some of the problems experienced possibly could have been alleviated if the families and hospitals had communicated earlier.

\textbf{6.65} Several witnesses commented that understaffing in hospitals also created a significant problem for both the families and the hospitals.\(^\text{466}\)

\textbf{6.66} The family of one vCJD patient arrived at the hospital one day to find that their son had not been fed. His medical notes stated ‘no-one available to feed’. Thereafter, the patient’s mother ensured that she attended the hospital in order to feed her son. The same family relied on people visiting other patients in the hospital to assist with the care of their son.\(^\text{467}\)

\textbf{6.67} In another example, a family asked nursing staff if their daughter could have more time to eat her meals as she was not able to cut up her food and experienced difficulty swallowing. The nurse apparently responded that the food trolley was only allowed on the ward for a certain amount of time in compliance with the occupational health and safety rules. The request for assistance to help cut up her daughter’s food was refused as the hospital was short-staffed.\(^\text{468}\)

\textbf{6.68} Neither of these two hospitals responded specifically to the allegations made by the families.

\textbf{6.69} A common symptom of vCJD sufferers was severe anxiety. A number of the patients experienced a terrible fear of hospitals\(^\text{469}\) and required constant care and reassurance while in hospital,\(^\text{470}\) an aspect of care that was difficult to accommodate in practice in busy hospitals.

\textbf{6.70} Generally, following referral to a neurologist, patients were sent to district hospitals for testing and assessment. Depending on where the patient lived, these hospitals could be relatively near, or far, from their home. Some patients’ families were upset that their relatives could not be given a bed near to their home. Being able to visit regularly was obviously important to the patients and their families.

\textbf{6.71} The experience of smaller community hospitals appears to have been largely positive. One witness said that she was ‘very impressed with the staff’ and had no problem with the care received.\(^\text{471}\) The fact that such hospitals tended to be nearer home was a definite advantage. It is clear that families appreciated the chance to assist in the care of their relatives while in hospital. Flexibility of staff and systems appear to have been an important issue in this respect.

\textbf{Communication and information}

\textbf{6.72} Communication and information appears to have been an issue of particular importance for several families. Being kept informed and having things explained to them appears to have been a priority in most cases. The importance of information was highlighted by one witness who said in her statement:

\(^{465}\) For example, S238 Carey response para. 4
\(^{466}\) S199 David Churchill paras 63–65; S571 Hodge para. 16; S596 Harrison para. 50; T72 p. 117
\(^{467}\) S199 David Churchill para. 64
\(^{468}\) S571 Hodge para. 16
\(^{469}\) For example, S569 Thorpe para. 32; S229 Sinnott para. 41
\(^{470}\) S229 Sinnott para. 41
\(^{471}\) T72 p.136
I would like to take this opportunity to especially thank the medical staff member of the National CJD Surveillance Unit, the medical and nursing staff at the hospitals at which [the patient] was nursed for their honesty, dedication and high standards of care. I have often been asked by people ‘Where did you get the strength to cope with the situation within which you found yourself?’ I believe that one does not know that one can handle such events until one is faced with them. However, my situation was very much assisted by the openness of the medical staff who kept me informed about [the patients’] illness from its very onset. I was never kept in the dark as to the progression of [the patients’] illness. This information-giving, I believe, resulted in reduced stress and an increased ability to cope with what was an extremely stressful situation.472

6.73 Several families were not happy with the level and manner of information provision, especially regarding the use of drugs and the reason for certain tests such as the brain biopsy.473

6.74 A report published jointly by the University of Edinburgh and the CJDSU in February 1999 found that several families also reported that they were informed of the diagnosis in an insensitive way.474

6.75 Several witnesses felt that information about a possible diagnosis was withheld from them. One stated that:

If we go back to the time of diagnosis at Queen Square, we never actually came away from that hospital with a specific diagnosis. The only clue that we received was when we saw our son’s brain biopsy operating notes which had in the top right-handed corner ‘Reason for operation’ and it had ‘CJD’.475

6.76 The same witness went on to explain how information was sought from various different doctors through the illness:

We contacted the GP initially and [the patient] was dismissed there. Then he went to the psychiatrist and was admitted to the psychiatric hospital there and there was nothing forthcoming there. Then he was admitted to the neurological department and there was nothing forthcoming there. So all the places we would have hoped there would be some information given to us there was nothing forthcoming. We were kept very much in the dark and the situation was not discussed with us.476

6.77 Other families had similar experiences, and found it difficult to find any information about CJD, even after the condition had been diagnosed. One recounted how she had resorted to going to the library and reading papers on the subject.477 Later on, especially with the development of support groups (see paragraphs 6.187–6.195), information provision improved dramatically.
An issue raised by several witnesses was that of dissemination of information about the disease within the medical community.\textsuperscript{478} As discussed earlier, lack of knowledge and awareness of CJD had many knock-on effects. Networking between hospitals was described as severely lacking,\textsuperscript{479} and it is possible that this may have led to delays in diagnosis, inappropriate care and further uncertainty for the relatives of vCJD victims. One witness also stated that she believed that the discovery of the new disease could have been moved forward by three months had communication between hospital doctors and the CJDSU been better.\textsuperscript{480} However, two neurologists who spoke at the Inquiry disputed this, and stated that dissemination of information was quick after March 1996.\textsuperscript{481}

**Involvement of the national Creutzfeldt-Jakob Disease Surveillance Unit**

As outlined in Chapter 4, any suspected cases of CJD were referred to the CJDSU through the neurological network. Each patient and their family was visited by a member of the CJDSU team in order to verify the diagnosis and obtain clinical and epidemiological information by way of a standard questionnaire.

The role of the CJDSU was another area which does not appear to have been properly explained to relatives, or why it was asking some of the questions it did about the victims’ medical history, eating habits, etc.\textsuperscript{482} Again lack of awareness of the disease had obvious implications for provision of information to the vCJD families.

One family complained that they felt that the doctor from the CJDSU expected them to take in too much information in a short space of time. They felt that he was more interested in obtaining permission from them for their daughter to undertake tests for research than in devising a care plan, or in assessing how the disease was affecting the family.\textsuperscript{483}

**Treatment and care of the patient following diagnosis of probable vCJD**

Sadly, vCJD is a terminal disease. Therefore, once a probable diagnosis of vCJD is made,\textsuperscript{484} the best that can be offered to a patient to date is palliative care. At this point families have to decide whether they wish to care for their family member at home, or whether that care should take place in a hospital or hospice.

**Caring for patients at home**

Many of the families of vCJD victims chose to nurse their family members at home.

\textsuperscript{478} T72 pp. 70–1  
\textsuperscript{479} T73 pp. 20–2  
\textsuperscript{480} T73 p. 71  
\textsuperscript{481} T72 pp. 39–40  
\textsuperscript{482} T73 p. 16  
\textsuperscript{483} S571 Hodge para. 24  
\textsuperscript{484} A diagnosis of vCJD is only able to be confirmed by post-mortem, therefore until post-mortem occurs a probable diagnosis of vCJD
6.84 The families who chose to do so tended to feel that:

i. they could provide a better level of care at home than their family member received in hospital; or
ii. they felt that their family member would be more comfortable at home; or
iii. they could not bear to see anyone but themselves care for their family member. 485

6.85 The families who chose to care for their family member at home tended to be the parents of younger victims of vCJD. 486 In many instances, this was only possible because at least one parent was able to give up work to care for their child full time, or one or both parents were fortunate to have sympathetic employers who enabled them to work flexible hours, to work part-time, to work at home, or to take extended leave without pay. 487

6.86 From a practical point of view, a number of the families have pointed out that they would not have been able to cope with caring for a child or spouse on their own because they would not have been able to lift them. A significant amount of lifting is required in the care of a vCJD patient, particularly as the disease progresses and mobility decreases. Patients must be lifted, for example, in and out of bed; from wheelchairs to commodes; in and out of the bath; and up and down stairs. 488

6.87 The evidence provided to the Inquiry has therefore shown that families who cared for their family members at home were unable to do so fully on their own. The statements have indicated that effective care required, among other things:

i. good support from the GP and other health professionals;
ii. the timely and appropriate provision of aids and equipment;
iii. financial support; and
iv. availability of respite for the carers.

6.88 One family was able to care for the patient throughout the illness, with hospitalisation only necessary at the time of their biopsy. 489 This case benefited from a supportive GP, good equipment and help from an occupational therapist, a district nurse and carers from a local hospice, illustrating the high level of care required for such treatment at home.

6.89 The issues surrounding the treatment and care of vCJD patients at home are discussed in greater detail below.

Provision of aids and equipment

6.90 Aids and equipment for vCJD victims were a very important feature of the care of patients at home and made a significant impact on their quality of life.

485 S205 Hassan para. 34; S204 Hall para. 26
486 For example, S214 Beyless
487 S224 Minto para. 22; S601 Stableford para. 44; S569 Thorpe paras 38 and 47
488 S601 Stableford paras 39–40
489 T73 pp. 3–4
6.91 Examples of very useful and vitally important equipment included the following:

i. Walking frames, wheelchairs, hoists, handrails, ramps and stair climbing apparatus for use as the mobility of patients decreased.

ii. Additional equipment including beds with sides to prevent falling, raised toilet seats, shower seats and air-cell mattresses.

iii. In the later stages of the disease items such as incontinence pads, suction machines, oxygen, syringes and gloves became indispensable.

6.92 Because of the severity of the disease and the fact that a patient’s condition can decline rapidly, prompt provision of appropriate beds and equipment is essential for adequate care. This appears to be one of the greatest areas of difficulty in caring for vCJD patients at home.

6.93 One of the problems experienced was that in a number of instances aids and equipment arrived too late to be of assistance. For example, in one case a wheelchair arrived too late, by which stage the patient was completely bedridden.490

6.94 Another problem was the provision of inappropriate equipment. In these cases equipment seemed to have been provided with little thought about its practical use. For example, a walking stick and walking frame were provided to one patient despite the fact that she could not use them on her own.491 Another patient was provided with a Zimmer frame with wheels on the front legs. This became a dangerous piece of equipment due to the patient’s poor balance, causing her to fall on several occasions.492 In another example, a stair climbing machine was found to be too dangerous as the patient was unable to control her movements and would fall to one side and risked hitting her head on the wall.493

6.95 Finance to obtain aids and equipment posed a problem for a number of families. One family told how they ‘had to fight to get everything’. The family approached a number of charities for financial assistance without success. Eventually, a care package came together after the family went to the local and national media.494

6.96 Another family bought some aids and equipment themselves.495 Many other families could not do so, especially when such items were very expensive, and only useful for a short period of the rapidly advancing disease, for example wheelchairs.

6.97 In July 1998, Irwin Mitchell, the solicitors for the relatives of the vCJD patients, prepared a paper on the provision and costing of care in the community for vCJD patients. The paper included a section on equipment and aids that had been provided to the vCJD victims being cared for at home. They noted that:

The general comment that all families made was that aids and equipment were only provided at their own request. This of course requires a knowledge of available equipment. This information should have been offered by the

490 S569 Thorpe para. 42
491 S222 Mellowship para. 18
492 S571 Hodge para. 18
493 S601 Stableford para. 41
494 S222 Mellowship paras. 39 and 41
495 S571 Hodge para. 29
occupational therapist (as no doubt they do provide the information when asked) but it is our understanding that this was not the case. 496

6.98 Irwin Mitchell also criticised the length of time it took for the agencies to provide help for vCJD victims:

In our experience the average family home cannot accommodate a terminally-ill patient without being adapted . . . The average time it takes for agencies such as the Occupational Therapy Service, Department of Social Services and local district councils to assess the suitability of a patient’s home and to approve the appropriate adaptations is one year . . . This does not take into consideration the problems with funding. Once again, this bureaucratic delay could be avoided with sufficient and minimum will. 497

6.99 There also appeared to be an area of particular disparity between different health authorities. One family were told that it would take 6 months for planning permission to make their home suitable. 498 Another explained how, although they were treated well in one region, it was very different when they moved to another county. Here no aids were offered, and equipment took a long time to arrive. 499

6.100 However, there were also reports of excellent and rapid provision of equipment, with one patient having a wheelchair especially made for her, and a stairlift, a hoist for the bath and a bed elevator installed. This particular family said that they felt that ‘everything that could have been done was done to make [their] daughter’s final days as comfortable as possible’. 500

The role of social services

6.101 As mentioned above, social service departments were involved both with the provision of equipment, and with staff such as social workers.

6.102 Again, a number of families reported difficulties with their respective social services departments. One described how they were sent a social worker who had no knowledge of vCJD, followed by a nurse who did little more than provide them with the absolute basic sanitary aids. 501

6.103 Another family described the social services as being ‘entrenched in their rules’ 502 as well as inefficiencies and delays. The rapid onset of the disease and the severity of its symptoms means that it was impractical for families to have to wait for equipment or to wait (up to 8 months in one case) 503 to be means tested for equipment.

6.104 On the other hand, several families indicated that they appreciated the assistance the social services gave in informing the family what benefits they could claim. 504

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496 S235 Provision of Care Report, p. 10
497 S235 Provision of Care Report, p. 10
498 T72 S235 Provision of Care Report p. 60
499 S215 Carey para. 40
500 S205 Hassan paras 29 and 30
501 T27 p. 124
502 S214 Beyless para. 37
503 S601 Stableford para. 45
504 S87 Keleghar para. 36. See also S205 Hassan para. 30
6.105 In the case of some of the earlier sufferers of vCJD, the families complained that they were either not offered help from social services, or there was no social services policy about how to treat vCJD patients, or the social services department did not know how to help and referred the family elsewhere.

6.106 As with the problems mentioned above regarding the provision of aids and equipment, it appears from statements provided by the families that a number of social services workers gave impractical advice relating to how to care for vCJD patients, or what aids and equipment to use.

The role of other healthcare professionals

6.107 A wide range of healthcare workers were also involved with caring for vCJD patients at home, including physiotherapists, district nurses, occupational therapists (OTs) and other specialised carers.

6.108 The statement of one OT provides a useful summary of the goals of the care team in one case. She wrote that the OT and the district nurse initially visited the family together to avoid duplication of work. They decided that the OT would be the most appropriate worker from the social services department to continue assisting the family. The OT visited to arrange and supply a number of aids and equipment. Financial issues were also discussed. The OT took part in a discussion with the family at the GP’s surgery to discuss the family member’s future needs. The OT said:

My aim within the team of workers was to be as unobtrusive to the family as possible. Much of what I did was preparation to be ready to respond as quickly as possible to [the patient’s] rapidly changing needs . . . I also anticipated what equipment he might need and had it ready for instant delivery, speed of response being so important in the handling of this condition.

She also noted that:

It was equally important to be able to remove the equipment as soon as it was no longer useful to avoid the distress of having it in the house.

6.109 She liaised with other workers by telephone and only visited when necessary as there were already many visitors to the house.

6.110 A number of families reported positive experiences and said that they had daily visits from home helps who ‘showed a professional and caring attitude’, as well as two or three visits per week from a district nurse. One family described how ‘an occupational therapist with the borough council arranged everything for us in terms of grants and equipment to help us nurse [the patient] at home’. Another family spoke very highly of their social worker who helped the family ‘through the
minefield of what we could and could not claim by way of benefits’. The social
worker kept in constant touch and remained in contact with the family after the
death of their son to ensure that the family was coping.\textsuperscript{514}

6.111 GPs also made a valuable contribution to the care of patients who were being
nursed at home. In many instances the GPs continued to support the patients and
their families. The families who praised their GPs in their witness statements
commonly referred to the attention and support that they received from the GP
during their entire ordeal. Often the support was as simple as visiting at home
regularly or reassuring the family that they were available for them 24 hours a
day.\textsuperscript{515}

6.112 Families also expressed gratitude for the efforts that their GPs went to to
organise and advocate for them for the provision of aids and equipment.\textsuperscript{516}

6.113 Families appreciated the availability of respite care, even if they did not use
the service.\textsuperscript{517}

6.114 Unfortunately, there were also families who reported a low level of support
from social services. One family told us that the district nurses were reluctant to
visit as they feared that CJD was contagious, and help was not offered by social
services or by the medical profession.\textsuperscript{518} Another family felt that the OT and social
worker were unable to cope and probably needed training in dealing with vCJD
patients.\textsuperscript{519}

\textbf{Care of patients in hospitals, hospices, disabled units and other
specialist hospitals}

6.115 A number of families were not able to care for their family member at home,
or decided against doing so. These families arranged for their family member to be
nursed in a hospital or hospice facility. Many of the families who did nurse their
family member at home eventually had to arrange for him or her to be cared for in
a hospital or hospice as the disease progressed and rendered their family member
increasingly incapacitated.

6.116 At this stage, the care was largely palliative and many families praised the
caring attitude of nursing staff and high standard of care.\textsuperscript{520} Good nursing care
removed a great deal of burden from the families, one of whom noted ‘The care
[the patient] received from the staff was magnificent. I could go home in the
evening with the knowledge that [the patient] would be well cared for’.\textsuperscript{521}

6.117 Many facilities were flexible enough to allow the patients to be made as
comfortable as possible. For example, one family said:

[The patient] had his own room where he could have his own things around
him, such as a television and his posters but most importantly the staff had
no objections to our being there all the time and to providing [the patient’s] nursing care.\textsuperscript{522}

Members of another family were able to stay overnight in the hospice in a spare bed.\textsuperscript{523}

\textbf{6.118} Specialist units where staff were used to dealing with long-term degenerative or terminal illnesses appear to have provided a much more appropriate level of care, both in terms of the patients themselves and the support for their families.

\textbf{6.119} Access to specialist facilities varied from region to region and not all patients were offered such care.

\textbf{6.120} Care in such institutions is obviously expensive, and in several cases there was debate over whether patients were actually entitled to receive treatment. One witness described how the chairman of the health authority had commented that ‘[the patient] is not a cancer patient, can she not be sent home?’\textsuperscript{524} Another described how their hospice had a policy that ‘an individual could only have a two-week stay unless . . . death was imminent’, although this particular patient was able to stay eventually.\textsuperscript{525} The uncertainty over whether the patient would have to be moved caused obvious anxiety. Another informed the Inquiry that the owner of the nursing home where they had placed their son was aware that there was an extensive need for more disabled and terminal nursing units for the young. The owner had discussed the matter with DH but, there being ‘no perception [within the Department] of a need for this type of unit’, had met with ‘brick walls’.\textsuperscript{526}

\textbf{6.121} Indeed, several families mentioned the difficulty of finding a long-term care facility suitable for a young person dying of a terminal illness.\textsuperscript{527} One of the families decided to nurse their child at home rather than in a home for the elderly.\textsuperscript{528} The hospice involved agreed that some aspects of its service were occasionally not suitable for some individuals, both elderly and younger in age. In the case of a younger person, the hospice advised that it was able to provide support in the patient’s home also.\textsuperscript{529}

\textbf{Financial costs to the families}

\textbf{6.122} CJD is a degenerative disease and as the disease progresses, the patient becomes more and more disabled, and requires an increase in the level of care. This imposes increased costs on families and breadwinners. Costs not only arise through the provision of care but also through miscellaneous expenses such as the purchase of household appliances, travel costs and increased household bills.

\textbf{6.123} DH’s Economics and Operational Research Division have estimated the cost of caring for vCJD patients. Based on information on patterns of care given in witness statements to the Inquiry, they estimated that care costs ranged widely, from under £6,500 to over £40,000. Their estimated cost per patient was £20,288.
However, the number of patients whose care was examined by DH was small and they were also unable to cost some aspects of care.530

6.124 Solicitors acting for the families of vCJD patients have put forward a statement to the Inquiry on the provision of care. They estimate that the cost of family care ranges between £39,500 and £45,500 for each family per annum. The cost represented the time spent on care by family members calculated on the basis of care assistant pay rates.531 We are not in a position to resolve the differences between these two estimates and have made no attempt to do so.

Perceptions of the risk to health care professionals

6.125 One area that caused the families considerable distress was the way that vCJD patients were treated with respect to the risk to medical and care staff. Again lack of awareness of human TSEs led to misunderstandings and difficulties. The risks involved and the sources of information available on this topic are discussed in detail in Chapter 8 of vol. 6: Human Health, 1989–96.

Post-mortem and burial/cremation issues

6.126 Following the deaths of their family members, the families quickly had to come to terms with a number of issues including burial or cremation procedures, death certification and inquests in England, Wales and Northern Ireland or fatal accident inquiries in Scotland.

6.127 Many problems appeared to have been encountered after death. Several families highlighted the distress caused by the suggestion that their loved ones had suddenly become an infectious risk to be handled while wearing protective clothing, when prior to death they had been nursing them without using any form of barrier.

6.128 One family in particular had problems getting permission for a burial rather than a cremation,532 and described how promises made to them over several post-mortem issues were broken.533 Several different parties were involved in the discussions and the decision-making process, including the coroner, the neurologist who carried out the autopsy, the vicar carrying out the funeral ceremony and the CJDSU.

6.129 Once the permission for a burial had been granted, the family were upset further by certain regulations that had been laid down regarding the burial procedures. These included the liming of the grave and body, the need for an especially deep grave, and the need for contractors to wear protective clothing. These were laid out in the City Council’s Ground Maintenance Contract Specifications, to be used for people who had died of infectious diseases. The family also said that they believed that public health officials were sent to the funeral to police these procedures.534

530 DH01 tab 25
531 S235 Provision of Care Report pp. 13–15
532 T73 pp. 39–40
533 T73 p. 33
534 T73 pp. 35–9
6.130 In a written statement the Honorary Secretary of the Coroners’ Society of England and Wales, Mr Michael Burgess, told us that:

My understanding is that coroners have no power to direct that any disposals must necessarily take any particular form or be accompanied by any special or abnormal hygiene arrangements – any such responsibility must lie with those responsible for local, public and environmental health matters.535

6.131 Professor Allen,536 the neurologist involved in the above case, has called for national guidelines on the burial of vCJD victims and the reporting of cases to the Coroner. This, she believed, was important due to the cultural/religious differences in burial customs throughout the United Kingdom. The matter was also discussed at SEAC’s 25th meeting on 8 March 1996.537 Dr Wight reported that DH had received a letter from the Northern Ireland CMO seeking the advice of the SEAC on funeral arrangements for CJD patients.

6.132 Problems were also encountered with autopsy arrangements. In one case 10 weeks passed before the post-mortem material was sent to the CJDSU, as the handlers refused to deliver it. No explanation was given, and eventually the CJDSU had to collect it themselves.538

6.133 In several cases, funeral directors and mortuary staff were also concerned about risks to their health. One witness explained that he had given permission for the body to be taken away for a post-mortem providing that he would be able to see it on its return. However, when the body was brought back it was in a body bag that the funeral director did not permit to be opened. The hospital had apparently stated that there would be no problem with opening the bag, but the undertaker had disagreed.539

6.134 Another witness was told by the undertaker not to touch her partner’s body because he had died of vCJD. The mortician had informed her that the reason why no one was allowed to touch him was because vCJD was an unknown disease and they did not know whether it could be passed via body fluid. In a written statement she said:

I then asked ‘Why were the children and I allowed to touch [the patient] when he was alive?’ He replied ‘You should not have touched him’.540

6.135 Some fear over infection was not unfounded, with information on how to handle bodies coming from hospital staff. One witness told the inquiry that:

When [the body] arrived at the family home we were instructed [by the undertaker] not to let young children into the room. We were instructed not to touch the contents of the coffin, not to touch [the body], not to touch the interior of the coffin.541

535 S637 Burgess para. 52
536 Professor Ingrid Allen was a member of SEAC from June 1990 to 4 December 1996
537 YB96/3.8/1.1–1.11 at 1.10
538 T73 p. 37
539 T73 pp. 41–4
540 S195 Beaney para. 58
541 T73 p. 34
The neurologist involved has since confirmed that she gave this advice because the skull had been opened during autopsy.  

### Death certification

The families of vCJD victims raised concerns about death certification. Many clearly indicated that the death certificates of their family members should record variant CJD as the cause of death. This issue was of particular relevance to some of the earliest known deaths from vCJD.

One family told the Inquiry about the problems caused by confusion over diagnosis, and the distress caused by not having a death certificate for several months after their son had actually been buried. They said:

> ... it just really compounded what we had been through, you know, previously, because things were still very much up in the air.

In the case of the earliest known death caused by vCJD, the death certificate recorded ‘bronchial pneumonia and progressive degenerative neurological disease’. Following post-mortem examination 6 weeks later it was concluded that the cause of death was vCJD. Eventually, after an inquest was held, the family was able to have the death certificate altered to record all three causes of death.

The following paragraphs examine the law and practice of death certification in England, Wales, Northern Ireland and Scotland.

### Registration of deaths: England and Wales

The relevant legislation governing the registration of deaths in England and Wales is the Births and Deaths Registration Act 1953 and the Registration of Births and Deaths Regulations 1987.

Section 22 of the Births and Deaths Registration Act 1953 provides that:

**22 Certificates of the cause of death**

(1) In the case of the death of any person who has been attended during his last illness by a registered medical practitioner, that practitioner shall sign a certificate in the prescribed form stating to the best of his knowledge and belief that the cause of death and shall forthwith deliver that certificate to the registrar.

(2) On signing a certificate of the cause of death under the foregoing subsection the medical practitioner shall give in the prescribed form to some qualified informant of the death notice in writing of the signing of the certificate, and that person shall, except where an inquest is held...
The problems of correct identification of the cause of death on the death certificate and of delay in correctly identifying the cause of death were discussed by the Office for National Statistics, which administers the law on the registration of births and deaths, in a memorandum to the Inquiry.

The memorandum explains:

When certifying a death, doctors are required to state the cause of death to the best of their knowledge and belief . . . However, in many cases of variant Creutzfeldt-Jakob disease, the diagnosis is made after death, and sometimes several months after the patient died. This is because the diagnosis of Creutzfeldt-Jakob disease has to be confirmed by neuropathological investigations. At the time of death of these patients the clinical picture is often that of a dementing illness of unknown aetiology. In such cases, variant Creutzfeldt-Jakob disease is only one of the differential diagnoses and it is usually not possible to be very certain about the precise cause of death until the results of post-mortem investigations become available.

The certificate completed by the medical practitioner is given to the registrar for the registration of the death. In certain circumstances the registrar must report the death to the coroner. Such circumstances are prescribed by Regulation 41 of the Registration of Births and Deaths Regulations 1987. These include, for example, where the cause of death appears to be unknown or the registrar has reason to believe that the cause of death was ‘unnatural’ (see below).

If the death is reported to the coroner, the registrar must await the outcome of the investigation or inquest before registering the death. In some situations the registrar may have recorded the death already, which is where complications may arise.

If no investigation, inquest or post-mortem is to be held the registrar must register the death according to the cause of death stated on the medical certificate within 5 days.

If a death is reported and he or she decided not to hold an inquest, then:

i. if a coroner’s post-mortem has been undertaken, the coroner’s notification is recorded in the register as the cause of death;

ii. if there has not been a coroner’s post-mortem, the information on the medical certificate is recorded as the cause of death.
6.149 If an inquest is held the registrar must register the death according to the findings of the inquest pursuant to section 23 of the Births and Deaths Registration Act 1953.\textsuperscript{555}

6.150 Section 23 of the Act covers the furnishing of information from the coroner and essentially provides the protocol for reregistering a death after an inquest.

Where an inquest is held into the death and the registrar receives under section 11(7) of the Coroners Act 1988 a certificate under the coroner’s hand –

a. giving information concerning the death; and

b. specifying the finding with respect to the particulars required to be registered concerning the death and with respect to the cause of death, the registrar shall in the prescribed form and manner register the death and the particulars found at the inquest, and, if the death has been previously registered, the said particulars shall be entered in the prescribed manner without any alteration of the original entry.\textsuperscript{556}

6.151 The prescribed manner for correcting of entries is laid down under section 29(3) of the Births and Deaths Registration Act 1953. This provides that:

(3) An error of fact or substance in any such register may be corrected by entry in the margin (without any alteration of the original entry) by the officer having the custody of the register . . . upon production to him by that person of a statutory declaration setting forth the nature of the error and the true facts of the case made by two qualified informants of the birth or death with reference to which the error has been made, or in default of two qualified informants then by two credible persons having knowledge of the truth of the case.\textsuperscript{557}

6.152 However, where ‘. . . an error of fact or substance occurs in the information given by a coroner’s certificate concerning . . . a death touching which he has held an inquest’, you may not change the details of the cause of death.

6.153 In essence therefore, if a post-mortem examination is carried out by request of the coroner, if the cause of death has already been registered, this may be changed by marginalia on the certificate. After an inquest, the coroner’s certificate will provide evidence for a new registration of death, which notes that a previous registration exists. The new registration is also noted on the previous one. If however, an inquest has been held and the coroner’s certificate itself records an error of fact as to cause of death, this can not be corrected on the death certificate.

6.154 Sections 44–46 of the Registration of Births and Deaths Regulations 1987 re-emphasise these requirements.\textsuperscript{558}

6.155 The registration of deaths in Scotland and Northern Ireland is not notably different from the system in England and Wales.

\textsuperscript{555} D001 tab 17
\textsuperscript{556} D001 tab 17, s. 23
\textsuperscript{557} D001 tab 17, s. 29(3)
\textsuperscript{558} D001 tab 19
Inquests and fatal accident inquiries

6.156 The primary concern of the families of vCJD victims in respect of inquests and fatal accidents inquiries has been that there has been no consistent practice of holding inquests or fatal accident inquiries following death by vCJD.

6.157 It has been submitted that ‘the overwhelming desire amongst families of the deceased is that an inquest should take place’. 559

6.158 The problem is essentially convincing a coroner that, under the terms of the Coroners Act 1988, a death from vCJD constitutes grounds for a post-mortem examination or indeed an inquest.

6.159 The following paragraphs examine the concerns of the families and the law and practice of inquests and fatal accident inquiries in England, Wales, Scotland and Northern Ireland.

Inquests in England and Wales

6.160 The law on inquests in England and Wales is codified in the Coroners Act 1988 560 and the Coroners Rules 1984. 561

6.161 According to section 8(1) of the Coroners Act 1988, the coroner is to hold an inquest where:

. . . a coroner is informed that the body of a person (‘the deceased’) is lying within his district and there is reasonable cause to suspect that the deceased

(a) has died a violent or unnatural death;

(b) has died a sudden death of which the cause is unknown.

6.162 Not every death becomes the subject of an inquest. Only when the above criteria are satisfied is a coroner empowered to act.

6.163 The concern that the families of vCJD victims have in relation to inquests is that arguably the various coroners in England and Wales had not consistently decided whether or not to hold an inquest.

6.164 Mr Stephen Irwin QC, Counsel for the families, has provided us with an opinion, in which he says:

The feelings of families are quite clear: the overwhelming desire amongst families of the deceased is that an inquest should take place. 562

6.165 Mr Irwin’s conclusion from a consideration of the cases dealing with whether a death is ‘unnatural’ or not is that a common sense approach must be

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559 M11F tab 15 para. 2
560 D001 tab 15A
561 D001 tab 15B
562 M11F tab 15 para. 2
adopted according to the individual facts of the case. He considers that the ‘common sense’ decision can be taken in three ways:

a. if the death is ‘unnatural’ in common sense terms;
b. if the cause of death is extremely rare;
c. death from a natural cause may become ‘unnatural’ due to a human failure which caused the death.

6.166 In his view, vCJD falls into the first two categories above. His rationale is that the accepted wisdom is that vCJD is caused by eating BSE-infected beef; it is a disease that has crossed the species barrier and does not otherwise arise in the human population. It is ‘unnatural’ according to the dictionary definition as it is ‘not in accordance with the usual course of nature’ and it is ‘at variance with what is natural, usual or expected; unusual, strange’.

6.167 The Honorary Secretary of the Coroner’s Society of England and Wales, Mr Michael Burgess, submitted a statement to the Inquiry setting out a helpful description of the role of the coroner in England and Wales and the questions that each coroner must ask him or herself during the course of an inquest.

6.168 Firstly, where the terms of section 8(1) of the Coroners Act 1988 have been met, the coroner is thereafter empowered to take an investigative role. The coroner will probably direct that a pathologist conduct a post-mortem examination. Once the post-mortem has been completed the coroner will again consider whether the provisions of section 8(1) have been met.

6.169 If an inquest is convened, the coroner will call witnesses who are able to assist in establishing how, when and where the death came about. Any medical notes will be obtained if possible and doctors may be called to give evidence or provide a report.

6.170 Mr Burgess describes the task of the coroner as follows:

At the conclusion of every inquest, an Inquisition is completed which sets out those matters which have been found and include a short factual statement as to how the cause of death came about, followed by a conclusion (the popular verdict – the ‘verdict’ is the entire findings, including the full name, address and occupation, the medical cause of death and circumstances as to how the medical cause of death arose).

6.171 The Inquisition records the particulars in relation to the death insofar as they have been proved on the balance of probabilities.

6.172 In relation to the issue of whether vCJD constitutes an ‘unnatural death’ for the purposes of section 8(1) of the Coroners Act 1988, Mr Burgess qualifies that:
This question has taxed many since its first discovery . . . Coroners are not bound by the decisions of their colleagues and their conclusions may differ from those of their peers.  

6.173 The difficulty with vCJD from the point of view of the coroner is that:

By the very nature of the disease, the ongoing research and the conclusions which flow therefrom, there is not a steady single unequivocal and certain source of knowledge and understanding to which there can be general reference.

6.174 Mr Burgess does not believe that:

. . . merely because a cause of death is rare or unusual (or, conversely, frequent or ‘usual’) it should for that reason necessarily be regarded as ‘unnatural’.

6.175 Mr Burgess also considers that if vCJD is caused by eating BSE-infected beef, it does not necessarily follow that death resulting from vCJD is ‘unnatural’ as there are other diseases that also cross the species barrier. Furthermore, he comments that there are causation difficulties in relation to vCJD:

With an incubation period measured in years, it would be unlikely that a particular meal or event could authoritatively be linked to the development of the fatal condition.

6.176 Mr Burgess issued guidance to members of the Coroner’s Society of England and Wales in a letter dated 19 November 1999. He commented that an ‘unusual’ death is not necessarily an ‘unnatural’ death. What constitutes an unnatural death ‘may well vary from time to time as understanding and knowledge evolves’. He affirmed that cases must be considered on their individual merits and approached ‘in a pragmatic way’. He concluded that:

. . . the answer to the statutory question ‘how’ is to be confined to ‘how the cause of death arose’ rather than the wider question ‘in what broad circumstances did the deceased die’.

This limitation does not enable us, in the context of an inquest into a single vCJD death, to reach general conclusions concerning the way in which this condition may be passed on or present itself . . .

6.177 Mr Burgess comments too on the suitability of the current system in dealing with cases such as vCJD, and suggests two alternatives:

i. Concern is expressed at the trauma that the publicity that an inquest provokes for the families. It is therefore suggested that an amendment to the Coroners Rules 1984 be made to give the option of excluding the public from certain inquests.
ii. It is suggested that a more appropriate inquisitorial forum may be a confidential inquiry which could consider the issues more widely than an inquest allows.  

Inquests in Northern Ireland

6.178 The system of inquests in Northern Ireland differs from the English and Welsh system and is governed by the Coroners Act (Northern Ireland) 1959 and the Coroners (Practice & Procedure) Rules (Northern Ireland) 1963. 

6.179 Section 13 of the Coroners Act (Northern Ireland) 1959 states that:

. . . a coroner within whose district –

(a) a dead body is found; or

(b) an unexpected or unexplained death, or a death in suspicious circumstances or in any of the circumstances mentioned in section seven occurs;

may hold an inquest . . .

6.180 The ‘circumstances’ mentioned in section 7 include:

- as a result of violence or misadventure or by unfair means;
- as a result of negligence or misconduct or malpractice on the part of others;
- from any cause other than natural illness or disease for which he had been seen and treated by a registered medical practitioner within 28 days prior to his death;
- in such circumstances as may require investigation.

6.181 Section 7 makes it mandatory for a death arising in the above circumstances to be reported to a coroner. Whether the coroner decides to hold an inquest is another matter.

6.182 If a coroner in Northern Ireland concludes that death caused by vCJD is a death caused by ‘other than natural illness or disease’, the words ‘may hold an inquest’ in section 13 of the Coroners Act (Northern Ireland) 1959, mean that a coroner has a discretion as to whether to hold an inquest or not. He or she is not compelled by the legislation to do so.

6.183 This issue therefore becomes whether a death arising from vCJD constitutes a death from a cause ‘other than natural illness or disease’.

577 S637 Burgess para. 48
578 S637 Burgess para. 49
579 DN01 tab 15A
580 DN01 tab 15B
581 DN01 tab 15A
6.184 The opinion of Mr Irwin QC is that ‘variant CJD should be regarded as an unnatural illness or disease’ and therefore should be reported to a coroner and that ‘holding an inquest would be appropriate and should be done’.582

6.185 Mr John Leckey, HM Coroner for Greater Belfast, gave consideration to the matter and gave his own opinion as follows:

The view I have reached is that in light of the present developing state of scientific knowledge concerning BSE and Creutzfeldt-Jakob Disease, it would be premature for me to make a firm decision as to whether an inquest should always be held in relation to such deaths. I believe the proper course is for me to consider the need for an inquest each time such a death occurs within my district. That would involve considering the available medical and scientific evidence as well as any views expressed by the family of the deceased. I suspect that not all families will share the same view as to the need for an inquest and that the scientific and medical evidence available will be of variable usefulness.583

Fatal accidents inquiries in Scotland

6.186 The Fatal Accidents and Sudden Deaths Inquiry (Scotland) Act 1976 provides the statutory framework for fatal accidents inquiries in Scotland. Under section 1, the Lord Advocate may order an Inquiry where:

It appears to the Lord Advocate to be expedient in the public interest in the case of a death to which this paragraph applies that an inquiry under this Act should be held in to the circumstances of the death on the ground that it was sudden, suspicious or unexplained, or has occurred in circumstances such as to give rise to serious public concern. In which case: the procurator fiscal for the district with which the circumstances of the death and apply to the sheriff for the holding of any inquiry under this Act in to those circumstances.584

Once the application from the procurator fiscal has been received, section 3 requires the sheriff to hold an inquiry ‘as soon thereafter as is reasonably practicable’.

6.187 Mr Irwin QC summarised the position as follows:

In summary . . . the Lord Advocate has a broad discretion to hold a fatal accident inquiry where a death arises from circumstances which give rise to serious public concern and in exercising this discretion the Lord Advocate will consider whether the inquiry relates to matters prejudicial to the health and safety of the public, and will also consider views of the family. It appears to me, as someone not qualified to advise with authority on Scots law, that this discretion certainly would arise in the context of variant CJD. Clearly, in any individual case the Lord Advocate would exercise his discretion rationally but should be influenced by an expressed desire on the part of the family that an FAI should take place. It is to be hoped that the Lord Advocate would also respond if the Committee of Inquiry into BSE expressed a

582 M11F tab 16 para. 7
583 S636 Leckey para. 3
584 D001 tab 18, s. 1
Organisations set up for the families of vCJD victims

6.188 In recognition of the pain and anguish experienced by the families coping with CJD a number of groups have been set up. The structure and function of these groups are described below.

CJD Support Network

6.189 During 1993 and 1994, Dr Will had several meetings with representatives of the Alzheimer’s Disease Society, in order to discuss the possibility of setting up a support network for the families of patients with CJD. In May 1994, a meeting was held at the Alzheimer’s Disease Society in London and the aims and organisation of the CJD Support Network were decided. A booklet was produced which provided information to the relatives of CJD patients, which was based partly upon the original CJDSU booklet. This booklet has been updated several times in order to keep it up to date with scientific discoveries.

6.190 The CJD Surveillance Unit had made money available for relatives to attend meetings of the Support Network. In 1996, £5,000 was donated to the Support Network from the Unit.

6.191 Dr Will is co-opted as a member of the CJD Support Network Committee and members of the CJDSU have written for newsletters, been present at meetings and given presentations.

Human BSE Foundation

6.192 On 28 July 1995, the Churchill family attended a meeting of the CJD Support Network mentioned above. On 21 May 1995, Mr and Mrs Churchill had lost their son, Stephen, the first recorded victim of vCJD.

6.193 The Churchill family subsequently became involved with the group and supported the publication of the booklet on CJD and its distribution to each neurology and psychiatry department and teaching hospital across Britain. They also proposed, funded and staffed the first telephone helpline for CJD on behalf of the CJD Support Network.

6.194 During the following 12 months, further cases of vCJD were ascertained and families of the victims became involved with the CJD Support Network.

6.195 However, the Churchill family felt that since vCJD affected a different population from classical CJD, a separate support group was needed. On 6 June 1997, the families of vCJD victims set up a separate support group, initially called the nvCJD Families Association. This group became known as the Human BSE Foundation in December 1997.
Since that date, the Human BSE Foundation has supported families of vCJD victims and campaigned for care packages and financial support for families. It has also successfully campaigned for the BSE Inquiry.588

Discussion of diagnosis, treatment and care of vCJD victims

In this chapter we discuss the experience of families of victims in respect of patient care and management of the illness of their family members. Experience since 20 March 1996 sheds light on the problems faced by families and their doctors before that date. Early diagnosis of this disease has been and continues to be a major problem as the first symptoms are of depression and other non-specific psychiatric features. It is only when neurological signs appear, especially ataxia, that CJD can be considered among the possible diagnoses.

The only non-invasive test which has to date been found helpful in the investigation is magnetic resonance imaging (an MRI scan), in which the ‘pulvinar sign’ (high signal level in the thalamus of the brain) is present in 78 per cent of patients affected by vCJD. The test is very specific in that it is not present in other forms of CJD and other neurodegenerative disorders. In the earlier cases of vCJD this test was not available, and the diagnosis was not normally made before death, except in a few cases, where brain or tonsil at biopsy revealed deposits of PrPSc. After death the neuropathological findings were diagnostic.

To this day there is no simple blood or cerebrospinal fluid test which is sufficiently reliable to be used in the ante-mortem diagnosis of vCJD. Research towards an immunological test is currently under way (see vol. 2: Science) but is not yet suitable for clinical application.

Only palliative treatment of symptoms is currently possible. Further basic research is required and under way into the function of the prion protein and into the factors which lead to the changes in conformation of the PrP which are associated with the pathological changes and with its infectivity. The most promising outcome of this research so far has been the development of β-breaker peptides which can reverse the conformational change of the protease–resistant prion protein in the test tube (see vol. 2: Science). If such a change can be induced by drugs in vivo, some hope may be afforded to patients in the early stages of disease. However, this will depend on the development of a reliable early biochemical test.

This chapter provides an account of the management and care provided to patients with vCJD and the level of support provided to their families. It is evident that in many cases this fell well below the standard that was desirable or would ordinarily be expected from the NHS and social services. The statements provided to the Inquiry by the families describe repeated shortcomings, misunderstandings and failures to appreciate the needs of young patients. In saying this we are not criticising individuals, but draw attention to failures in the system. In a number of instances the families praise the care and attention they received from medical and nursing staff. These instances are important in that they illustrate the level of care

588 S231 Human BSE Foundation paras 21–6
that can often help to mitigate the stress and pain of the victims and relieve the anxiety of family carers.

**6.202** These shortcomings do not appear to be due to lack of resources, but rather to a failure to appreciate the needs of young patients dying of a progressive neurological illness. Variant CJD emerged as a new and complex disease about which little was known. Staff were seeing the condition for the first time and had little to guide them on prognosis and management. When it was appreciated that aids such as wheelchairs and bathroom lifts were required, they often arrived when they were no longer of use to the patient.

**6.203** In many instances, the day-to-day care of the patient was left in the hands of the family members, who often had to leave their employment to devote time to nurse the patient 24 hours a day. It should be emphasised that families preferred to look after the patient at home, but this was associated with a considerable physical and financial burden for which the families received insufficient help. In several instances, respite care was provided for family carers but was not made available to others. This is only one example of the inequality in provision that existed throughout the country.

**6.204** Mention should be made of the support to families provided by the Human BSE Foundation and the CJD Support Network. The Foundation is a support group formed by the families of vCJD victims. They distribute literature and provide a telephone helpline. Their personal experience has been used in many ways to improve the care given to patients.

**6.205** It seems that patients for whom a care plan had been carefully arranged have received better management than those for whom this was lacking. It should be possible for all suspected vCJD cases to receive such a care plan now that a care coordinator has been appointed by the CJDSU to provide advice to statutory care providers in both the health and social services sectors and to act as an advocate on behalf of patients with vCJD and their families.