CJD SUPPORT NETWORK

HELPLINE 01630 673973

Newsletter

Issue 11

November 2002

CREUTZFELDT-JAKOB DISEASE SUPPORT NETWORK NEWSLETTER

Chief medical officer welcomes vCJD research results

Chief medical officer Sir Liam Donaldson welcomed the publication, in the British Medical Journal (BMJ) on 20 September, of preliminary results from an extensive survey, showing abnormal prion protein in a single appendix. Sir Liam stressed the uncertainties of the implications of this finding. He announced plans to set up a new archive of tonsils for more research to support a better estimate of the prevalence of vCJD.

The published results are from the second phase of a study of specimens from over 8,300 patients aged between ten and 50, who had appendicectomies and tonsillectomies for reasons unrelated to CJD. The study is looking for evidence of abnormal prion protein that could be a marker for vCJD. The first phase of this study was published in April 2000 and found no positive samples. This second phase of the study is due for completion in 2003.

It is difficult to know the public health significance of a single positive finding of prion protein. Using this single positive result to predict the number of individuals who might also test positive results in wide margins of error. This prediction ranges from 0.5 to 900 people per million in the age group tested – a level of statistical uncertainty in line with existing statistical models, which have calculated a range from a few hundred to over 100,000 cases. The long incubation period of the disease means we cannot yet have any firm idea of how many people will eventually get vCJD.

Sir Liam said, 'There is still so much that we don't know about variant CJD. This research provides evidence of infection, but it is very hard to draw conclusions on the size of the outbreak from it. We need to continue to increase our understanding of the disease. The studies reported in the BMJ are continuing and another study on tonsils is ongoing at the MRC Prion Unit in London.

'More research is needed. The steering group for these research studies, chaired by Professor Sir Leszek Borysiewicz, has advised me that it is important to collect as large a number of samples as possible, and we are now planning to set up a new tonsil archive.

'This further research is important for scientific and health service planning purposes. Public health action in this field is undertaken on a precautionary basis and is not dependent on awaiting the results of long term studies.

'The steering group has given much thought to the ethical issues associated with a collection of this sort. They have recommended that this be an anonymous archive, which means that individual patients will not be identified from their tonsil samples. The archive will allow prospective studies to be done on the largest possible number of tonsils, so that we can get better estimates of the size of the vCJD epidemic.'

International CJD Day

The first International CJD Day takes place on Tuesday 12 November 2002. A one-day conference, 'Aspects and Perspectives', will be held in Liverpool, with linked events taking place on and after that date.

12 November

- Conference (Liverpool)
- Launch of nursing guidelines by the CJD Support Network and the Queen's Nursing Institute
- Launch of patients' and carers' guide by Brain & Spine Foundation

16 November

- Concert in memory of Mrs Midwinter (Lancaster)

17 November

- CJD memorial service (London)

See page 2-3 for details

ON OTHER PAGES

Published by Alzheimer's Society, Gordon House, 10 Greencoat Place, London SW1P 1PH. Tel 020 7306 0606. Email cjdnet@alzheimers.org.uk. Website www.alzheimers.org.uk
Registered charity no. 296645. A company limited by guarantee and registered in England no. 2115499

Aspects and Perspectives

A one-day conference on CJD

12 November 2002 (International CJD Day)

9.30am to 4.15pm · £85.00 (including refreshments)

Glaxo Neurological Centre, Liverpool L3 8LR

SPEAKERS

- Dr Angus Kennedy, consultant neurologist, National Prion Clinic: *What is CJD?*
- Professor JW Ironside, consultant neuropathologist and director of the National CJD Surveillance Unit: *What controls are in place or are needed?*
- Dr M Doran, consultant neurologist, Walton Centre
- Dr Larner, consultant neurologist, Walton Centre: *A local perspective*
- Professor Don Jeffries, vice president of the Royal College of Pathologists and vice chair of the CJD Incidents Panel: *Infection control*
- Mr R Tomkins and Ms S Shadbolt: *Carers' experiences of vCJD and sporadic CJD*

Chair: Gillian Turner, CJD co-ordinator, CJD Support Network

This event will be of interest to a wide range of people involved in the care and support of those affected by CJD

Balloon launch in Liverpool

Hundreds of balloons will be released in Liverpool at 1.30pm on 12 November in memory of the people who have died of CJD in the last ten years. The balloons will be released by Zoey Appleyard, whose aunt, Baroness Wharton, died of sporadic CJD. Of the 708 balloons to be released, 502 yellow ones will be in memory of those who died of sporadic CJD, 34 green ones for those with iatrogenic CJD, 50 white ones for those with familial CJD and 117 red balloons for those with vCJD.

CJD Support Network to launch nursing guidelines for CJD

On 12 November (to mark International CJD Day) the CJD Support Network, in conjunction with the Queen's Nursing Institute, is launching nursing guidelines for CJD.

The guidelines are aimed at nurses who are caring for patients with all types of CJD. They have been developed with the help of people who have cared for family members with all types of CJD, and professionals from a wide range of care disciplines. The project has been funded by the Department of Health.

A copy of the guidelines can be obtained from Gillian Turner, co-ordinator of the CJD Support Network.

Launch of Nina Nadine rose

A Florabunda bush rose was presented this year at Hampton Court flower show. It is named after Nina, a young girl who died of vCJD in 1997, and it is in memory of all the young people who have died of this terrible disease. It is a beautiful colour, beginning as yellow, tinged with pink tips. As the rose blooms, the colour changes to a soft pink; it also has a gentle fragrance.

Pene and Mike Sinnot will launch the rose on International CJD Day at the conference at the Glaxo Neurologist Centre.

The Nina Nadine rose is available at a cost of £7.50 from:
Cand K Jones, Golden Field Nursery, Barrow Lane, Tarvin, Cheshire CH3 8JF
Telephone 01829 740663
Fax 0182741877

Brain & Spine Foundation to launch CJD information

Not only does 12 November 2002 mark International CJD Day, but it also marks the launch of a package of CJD information tailored specifically to the needs of families, as well as the GPs that care for them.

CJD – a guide for patients and carers is an illustrated 32-page booklet that is being published by the Brain & Spine Foundation together with a 12-page companion leaflet called *CJD – a guide for GPs*. The package provides essential information on what CJD is, the different types of CJD and what support and treatment is available. Surveys of affected families, carried out by the Brain & Spine Foundation and the CJD Support Network, highlighted a need for more information and practical advice from GPs. This included information on benefits available and where to get support and advocacy. Information about the impact of the illness on the rest of the family also ranked high on the list of concerns.

Maggie Alexander, director of the Foundation said, 'This leaflet has been produced in close consultation and collaboration with families and carers. We have taken on board their desire for clear information and also their concern about lack of CJD knowledge amongst GPs, and the booklet and leaflet reflect this.'

Memorial concert

Saturday 16 November, Lancaster (Greaves) Methodist church, 7.30pm

Clifford Midwinter's wife died of sporadic CJD in October 2001. He has arranged a concert of light vocal and instrumental music, to be held in her memory at Lancaster (Greaves) Methodist church on 16 November.

If you would like to attend the concert, phone Clifford on 01524 33009 or write to him at 64 Newlands Road, Lancaster, LA1 4JF. The tickets cost £3 for adults and £2 for children.

CJD memorial service

Sunday 17 November, St Martins in the Field, Trafalgar Square, London

Since holding the first memorial service for those who have died of CJD, in December 1999, we have been repeatedly asked by members to hold another similar service.

To mark International CJD Day, we have arranged a memorial service to be held on Sunday 17 November at St Martins in the Field, Trafalgar Square, London at 6.30pm.

At the service, the names of people who have died from CJD will be read from a memorial roll and a candle will be lit in their memory. After the service, there will be an opportunity to meet other families over a cup of coffee in the crypt.

News in brief

Medical Research Council

The Medical Research Council hosted a consumer workshop on clinical trials for CJD on 26 July. This was an excellent day and was attended by many members of the CJD Support Network.

CJD therapy group

The chief medical officer has set up a CJD therapy group to advise the Department of Health on its response to emerging drug therapies for CJD that may be suitable for clinical trials. The group will be chaired by Professor Sir Michael Rawlins and will include Lester Firkins of the Human BSE Foundation and Gillian Turner of the CJD Support Network.

vCJD Trust

The vCJD Trust has been established, and trustees appointed by the secretary of state, to assess compensation claims from families affected by vCJD. Charles Russell, Solicitors have been appointed secretariat and will advise the trustees in relation to claims. To obtain relevant forms and details please contact Edwina Rawson on 020 7203 5335.

Inquiry launched after rise in tonsil operation deaths

The Times reported on 9 August 2002 that a major audit of all tonsil operations has been started to establish if disposable surgical instruments, introduced because of the BSE epidemic, caused a rise in deaths and complications.

British blood too risky for our children

The Daily Mail reported on 16 August 2002 that hospitals are to import US blood supplies to ensure that children under six are not infected with vCJD. This follows research suggesting that the agent that causes vCJD is transmitted much more easily than had been thought.

Onset of vCJD in the first 100 cases

Michael D Spencer, Richard SG Knight and Robert G Will published a paper in the British Medical Journal, 22 June 2002; 324:1479-1482, called *First hundred cases of variant CJD: retrospective case note review of early psychiatric and neurological features*.

Research results showed that the early stages of vCJD are dominated by psychiatric symptoms, but neurological symptoms precede psychiatric symptoms in 15 per cent of cases. They are present in combination with psychiatric symptoms in 22 per cent of cases, from the onset of disease. Common early psychiatric features include dysphoria, withdrawal, anxiety, insomnia, and loss of interest.

No common early neurological features exist, but a significant proportion of patients exhibit neurological symptoms within four months of clinical onset, including poor memory, pain, sensory symptoms, unsteadiness of gait and dysarthria.

Their conclusion was that although the diagnosis of vCJD may be impossible in the early stages of the illness, particular combinations of psychiatric and neurological features may allow early diagnosis in an appreciable proportion of patients.

Creutzfeldt-Jakob disease surveillance in the UK

A summary of the tenth annual report of the National CJD Surveillance Unit

The national surveillance programme for CJD was initiated in 1990. In 1999, the National CJD Surveillance Unit (NCJDSU) became a World Health Organisation (WHO) collaborative centre for reference and research on the surveillance and epidemiology of human transmissible spongiform encephalopathies. The National Care Team, based within the NCJDSU, was formed in September 2001. The team consists of two care co-ordinators, a neurologist and a secretary.

The tenth annual report of the NCJDSU shows that it has obtained clinical and epidemiological information for most CJD patients. The case-control study for CJD has been extended for vCJD, with up to four community controls studied in addition to the hospital controls. The post mortem rate for patients with suspect CJD is high, although following the Alder Hey inquiry, there is evidence that autopsy rates have declined in line with autopsy rates in the UK generally.

Sporadic CJD

From 1990-2001 mortality rates from sporadic CJD in England, Scotland, Wales and Northern Ireland were, respectively, 0.77, 0.85, 1.03 and 0.52 per million a year. The difference between the rates in each country is not statistically significant. These rates are comparable to those observed in other countries in Europe and elsewhere in the world, including countries that are free of BSE. In England, the highest and lowest mortality rates from sporadic CJD were observed respectively in the South West (standard mortality ratio [SMR] = 129) and East and West Midlands regions (both SMR = 83). This variation is also not statistically significant.

Recent CJD figures

The number of suspect cases referred to the CJD Surveillance Unit in Edinburgh, and the number of deaths of definite and probable cases of vCJD in the UK, up to 4 October 2002:

Deaths	
Deaths from definite vCJD (confirmed):	93
Deaths from probable vCJD (without neuropathological confirmation):	24
Deaths from probable vCJD (neuropathological confirmation pending):	0
Number of deaths from definite or probable vCJD:	117
Alive	
Number of probable vCJD cases still alive:	11
Total number of definite or probable vCJD cases (dead and alive):	128

Variant CJD (vCJD)

Up to 31 December 2001, there were 104 deaths from definite or probable vCJD in the UK; of these, 89 were confirmed neuropathologically, with one additional case awaiting neuropathological confirmation. The clinical, neuropathological and epidemiological features of all these cases of vCJD are uniform and consistent with the NCJDSU's previous descriptions. Analysis from January 1994 to December 2001 shows evidence of an annual increase of around 20 per cent for both onsets and deaths, but because of the uncertainty over the incubation period of vCJD, continuing surveillance will be required to establish whether this trend is sustained in future years.

Risk factors

Risk factors for the development of vCJD include age, residence in the UK and methionine homozygosity at codon 129 of the prion protein gene (all 98 cases of vCJD with available genetic analysis have been methionine homozygotes). The analyses in this report do not provide conclusive evidence of an increased risk of vCJD associated with past surgery, previous blood transfusion, occupation, or a range of dietary factors. However, the effectiveness of the case-control study, from which these results are derived, is limited by the relatively small number of cases and controls.

Diet

Patients of vCJD are reported to have consumed products potentially containing mechanically recovered meat (such as sausages and burgers) more frequently than community controls. However, care should be taken in interpreting this result, as there is considerable scope for recall bias with respect to dietary history.

'North-South divide' and clusters

The incidence of CJD across the UK continues to be greater in the north of the country. The reason for this is not clear and further investigations are required to investigate the possible causes. The only statistically significant cluster of vCJD cases in the UK is in Leicestershire. Geographically associated cases of vCJD are subject to detailed investigation involving the NCJDSU, colleagues at the Communicable Disease Surveillance Centre (CDSC) and local public health physicians.

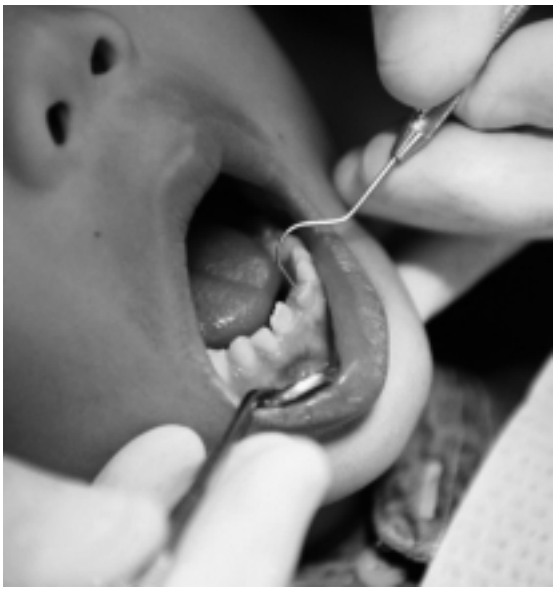
Collaboration and support

The NCJDSU reports that its activities are strengthened through collaboration with other surveillance projects, including the Transfusion Medicine Epidemiology Review and the Progressive Intellectual and Neurological Deterioration in Children Study. The collaboration of colleagues in these projects is greatly appreciated. The success of the NCJDSU continues to depend on the extraordinary level of co-operation from the neuroscience community and other medical and paramedical staff throughout the UK. The NCJDSU is particularly grateful to the relatives of patients for their help with this study.

The full NCJDSU report is available at: www.cjd.ed.ac.uk/rep2001.html

REFERRALS OF SUSPECT CJD		DEATHS OF DEFINITE AND PROBABLE CJD						
Year	Referrals	Year	Sporadic	Iatrogenic	Familial	GSS	vCJD	Total deaths
1990	[53]	1990	28	5	0	0	-	33
1991	75	1991	32	1	3	0	-	36
1992	96	1992	44	2	5	1	-	52
1993	78	1993	37	4	3	2	-	46
1994	116	1994	51	1	4	3	-	59
1995	87	1995	35	4	2	3	3	47
1996	134	1996	40	4	2	4	10	60
1997	161	1997	59	6	4	1	10	80
1998	154	1998	63	3	4	1	18	89
1999	169	1999	61	6	2	0	15	84
2000	178	2000	48	1	2	1	28	80
2001	173	2001	52	3	2	2	20	79
2002*	107	2002*	27	0	2	0	13	41
Total referrals	**1581**	**Total deaths**	**577**	**40**	**35**	**18**	**117**	**786**

*As at 4 October 2002



CJD and the dentist

Dr Andrew Smith and Dr Petrina Sweeney, senior lecturer in special needs dentistry, Glasgow Dental School

Early contact with dental services is strongly advised when a person has been diagnosed with CJD, to ensure maintenance of good oral and dental health. Any existing dental problems should be dealt with as early as possible to minimise invasive treatment and dental pain at a later stage in the disease. The provision of dental services for patients with special needs varies throughout the country, but the local community dental service should be contacted in the first instance.

A dental hygienist should be involved in the patient's care, to give advice and maintain regular oral hygiene, particularly if the patient has had advanced restorative work or if their level of impairment hinders satisfactory self-care. Special infection control measures are not required for dental treatment other than the disposal of instruments used by the dentist. For further advice, contact the CJD Support Network.

Once a comfortable, clean and healthy oral environment has been established, simple oral hygiene provided by family members and care-workers is all that is required to allow the patient to maintain normal function.

CJD palliative care research

Dr Emma Jones, St Thomas' hospital, London

I am a doctor working in palliative medicine (hospice work). Last year I was involved in the care of several patients with CJD, and I started asking questions about the symptoms and problems that these patients and their families experienced. I wanted to know the best way to alleviate their symptoms and to support the family, or at least to know how other healthcare professionals were dealing with these issues. Although there has been a lot of research into CJD, it has tended to focus on scientific areas rather than care issues.

My questions led me to develop two studies designed to inform health professionals and carers about the issues involved in the palliative care of a patient with CJD.

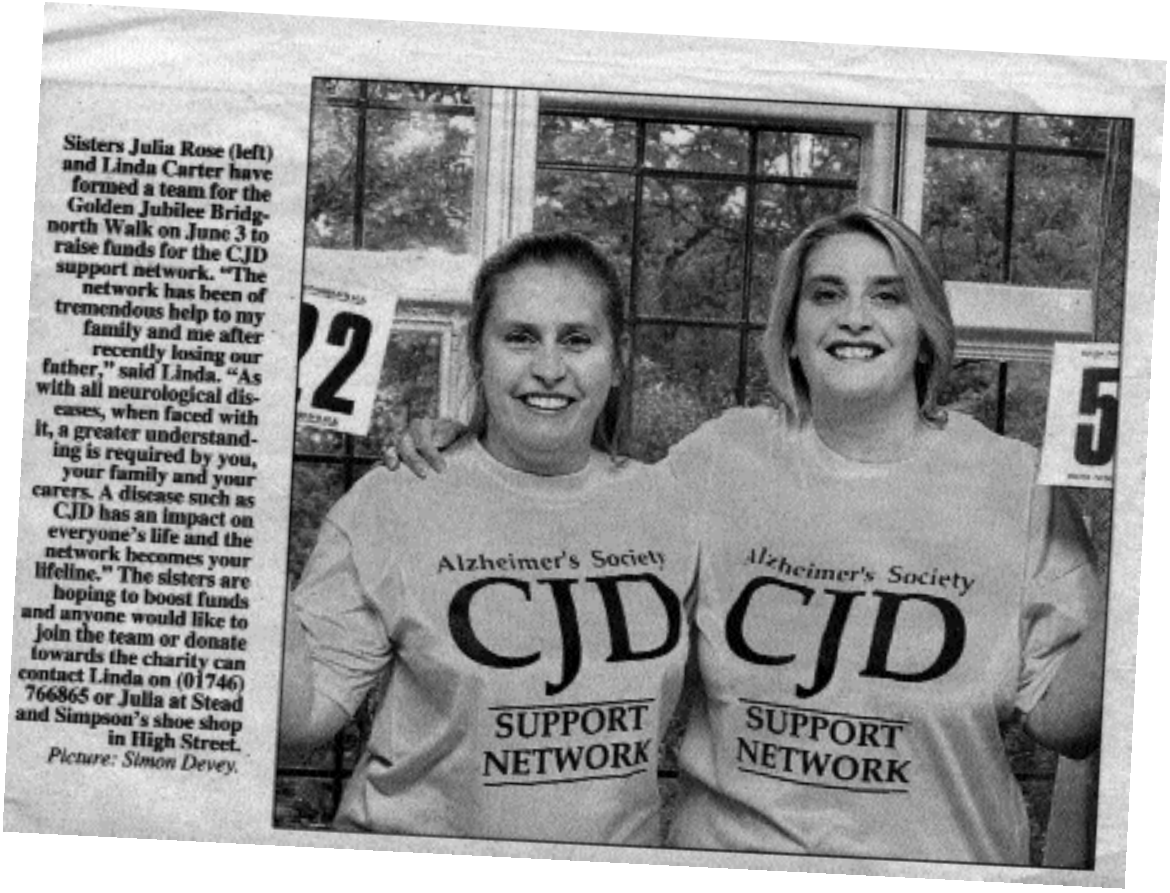
My first study was a nation-wide postal survey of all hospices and specialist palliative care services in the UK (652 in total). The questionnaire asked about referral policies and the experiences of patients with CJD, and invited respondents to participate in a telephone interview if they had recently looked after a patient with CJD. The response was overwhelming: 80 per cent replied and 110 people volunteered to be interviewed. I have selected 30 professionals who have recently looked after a patient with variant CJD and the interviewing has now started. All professionals and any patients they discuss will remain anonymous; the study has been given ethical approval from the South East multi-centre research ethics committee.

Hospital palliative care teams, hospices, day centres and community palliative care teams have all had a role in the care of patients with vCJD. I am asking some open questions about the issues encountered, so that respondents can describe problems and difficulties as well as things that have gone well. One respondent said that caring for a vCJD patient was 'uncharted territory', but that they had also discovered that, in many ways, it was, 'no different to treating anyone else in palliative medicine,' in that the multidisciplinary team approach, problem management and medications used were essentially the same. It will be interesting to see if that is a representative view.

Plans are under way for a follow-up study of patients with vCJD. This study will look at the burden of symptoms and problems facing patients, families and professionals over time.

I am grateful to the CJD Support Network for providing an information booklet to accompany the questionnaire. It was sent to all hospices and specialist palliative care services in the UK. I am also grateful to the Human BSE Foundation for their continued financial support.

Completing the Bridgnorth walk



Picture: Simon Devey/Bridgnorth Journal

Linda Carter

In November 2001, my father died of probable sporadic CJD. I had lots of contact with the CJD Support Network, initially from the website as I tried to find out more about the disease. I spoke with Gillian, the Support Network co-ordinator, later on as my father's disease progressed. My family and I had great support from her during the many difficult times of need. The Support Network gave us the best understanding of CJD possible.

Our father spent his last few days close to home at a local hospice. My brother, my sister and I met Gillian there to discuss our concerns as a family. After our father passed away, my sister and I decided that we wanted to raise awareness of the CJD Support Network and to raise money for it at the same time – we also wanted to make our father proud.

Our father had always been a keen walker and a member of local rambling groups in Shropshire, and campaigned to keep footpaths open. We decided, therefore, to try and complete the annual Bridgnorth walk of 22 miles over a series of hills (very big hills!)

We managed to gather a team of six people: my sister Julia Rose; her son Glenn Humphries; Arthur Beyless, treasurer of the CJD Support Network, who lost his daughter Pamela to vCJD; Helen Slater; Paul Brettle, and me.

The walk went well; it rained the whole way, apart from the last four miles – so then we had a welcome chance to dry off! Many people walked this year – there were about 800 people, supporting many different charities. I think we had the best T-shirts, sporting the name of the CJD Support Network.

We raised about £500 as well as fantastic awareness for the Network, which was well deserved. Our father would have been proud of what we achieved. We only had the strength to do it because of him. Hopefully, his positive outlook will inspire others to raise awareness in the future.

Heart rate variability as an aid to diagnosis in vCJD

Laura Woolfson, research associate, University of Manchester

Although much research has been done into diagnosing vCJD, the only tests available at present to determine a diagnosis are a tonsil biopsy or a brain biopsy, both of which involve surgery and a stay in hospital. Early diagnosis is essential, so that when treatments are found they can be started soon after the onset of symptoms.

However, a simple test for vCJD that can be done in your own home is currently being researched at the University of Manchester. I am a research associate in the department of anaesthesia at the university. Our team hopes to use changes in heart rate variability as an aid to the diagnosis of vCJD. To do this, we are using a medical monitor called Fathom™ (Amtec Medical Ltd) This was co-invented by one of the team, Dr Chris Pomfrett. Fathom has been used for the last ten years to determine the depth of sedation in patients during surgery.

Fathom works by measuring respiratory sinus arrhythmia (RSA). RSA is a normal change in heart rate, linked with breathing. Areas of the brain called brainstem nuclei control the timing of heartbeats. We believe that vCJD affects some of these brainstem nuclei, which in turn affects RSA. We are trying to show that vCJD causes a specific change in RSA that could help to distinguish it from other forms of the disease.

The test involves measuring the heart rate using four stick-on electrodes. The breathing rate is also measured. The data obtained is analysed by computer, and changes in heart rate variability that are not visible to the naked eye on a heart trace (ECG) are picked up.

I liaise with health professionals and organisations working with people who have CJD and their carers to raise awareness of this study, and I provide information to potential volunteers and interested parties. The research team is also grateful to Dr Richard Knight and the team at the National CJD Surveillance Unit in Edinburgh for their help in giving information sheets to patients referred to them.

If someone is interested in the study, I send them a detailed information sheet, which I follow up with a phone call. This enables me to talk in depth with the volunteer or their carer and to arrange a suitable time to visit. One of the many advantages of Fathom is that it is portable, so I can visit the volunteer at home. I am aware that it will not always be convenient for me to visit between 9am and 5pm and I am happy to visit 'out of hours' or at weekends if this is more convenient.

I would be happy to discuss the study further, or to answer any questions you may have about the research. You can contact me on 0161 2765012.

Tonsil biopsy in vCJD disease

Professor John Collinge MD FRCP FRCPath FmedSci, consultant neurologist at St Mary's Prion Clinic

There are a number of forms of CJD but only one of these, variant CJD or vCJD, is linked to exposure to bovine spongiform encephalopathy (BSE). Around 120 people are confirmed as having the disease in the UK (March 2002). However, because the incubation periods of these diseases in humans are extremely variable and long it remains possible that a substantial epidemic of vCJD will occur in the years ahead.

Difficulty of diagnosis

Diagnosis of vCJD can be difficult, particularly in its early stages when the disease resembles common psychiatric conditions such as depression. However, it is very important to reach a diagnosis, as there are a number of other diseases that cause similar symptoms to vCJD, and some of these respond to treatment. In some patients, the features of the disease and the various tests performed may make a diagnosis of vCJD very likely; in others, the situation may be less clear. For this reason, a brain biopsy may be considered.

Why a tonsil biopsy?

It has been known for many years that prion diseases in animals involve other parts of the body as well as the brain. For example, the lymphoreticular system (LRS),

which is part of the body's defence against infection, becomes infected with prions at a very early stage. Instead of defending the body, the LRS acts as a place where prions can 'breed' and infect other parts of the body, such as the brain. The LRS includes the tonsils. By removing a small piece of tonsil tissue in appropriate patients, it is possible to detect the rogue form of prion protein that is thought to be the cause of CJD. This can confirm the diagnosis so that other tests, including a brain biopsy, are not needed. The tonsil test is only positive in vCJD and not in other forms of CJD.

What does the test involve?

At present, tonsil biopsy is only performed at the National Prion Clinic at St Mary's hospital, London, which has specialised facilities for patients affected by prion diseases. Patients are normally referred to this service by consultant neurologists, and the patients will usually have already undergone extensive testing at their local hospital and perhaps at a neurological centre.

The assessment at St Mary's is planned over a three- to five-day period. On the first day the patient is examined by doctors, to consider whether a tonsil biopsy will help. The diagnosis and recommendations for further tests are then discussed with the patient and family, and a plan of action agreed with them. If a tonsil biopsy is recommended, it is then performed. Some patients will have had their tonsils removed during childhood but usually sufficient tonsil remains to do a successful biopsy. The test requires a general anaesthetic, so the patient is assessed by an anaesthetist as well as by the ear, nose and throat surgeon who performs the biopsy.

The biopsy itself involves removing a small piece of tissue from one of the two tonsils; it is a very simple procedure. A special disposable biopsy kit is used. The patient normally stays in hospital overnight to ensure all is well, before returning to their local hospital or home the following day.

St Mary's is a busy teaching hospital, and there are often pressures on availability of beds. The patient may, therefore, need to wait a little longer for a biopsy. Our staff appreciate that this is a very stressful time for patients and carers, and do everything they can to help things run smoothly and rapidly.

Once the biopsy has been taken, two separate types of test are performed. One involves looking at the tissue through a microscope after treating it with a special stain to detect the rogue form of the prion protein. The second test detects and characterises the rogue prion protein using a method called Western blotting. Both techniques are time-consuming and sometimes need to be repeated to get a clear result. Usually a result is available about one week after the biopsy and this is either communicated to the patient and/or family by their local doctor or during an out-patient visit to St Mary's.

Need to know

It is very important to make a diagnosis to be sure that no other disease, which may respond to treatment, is causing the condition. Although devastating, a firm diagnosis of vCJD ends uncertainty and removes the need for any further tests, including brain biopsy. Establishing a diagnosis is also extremely important in developing an individualised care package for each patient. If the tonsil biopsy is negative, other tests may be advised.

While tonsil biopsy is a relatively simple procedure, it does involve a small operation and an anaesthetic. The ideal would be to develop a blood test to make the diagnosis. So far this has not proved possible but much research is underway by us and others to try to develop such a test.

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Inquests

Dear Sirs,

My daughter Pamela died from vCJD. Since her death in October 1998, I have, like many other families, fought long and hard for an inquest into her death.

A question often asked about the tragedy of vCJD is: why an inquest? My campaign, *Action for Inquests* will attempt to answer that question.

The purpose of an inquest is to examine the circumstances around the death of an individual that is reported as being suspicious, violent or unnatural (section 8(1) of the Coroners' Act, 1988).

Reported *to* whom? The coroner. The coroner is an independent judicial officer who must follow the laws that apply to coroners and inquests.

Reported *by* whom? The GP, hospital or registrar of births, deaths and marriages. Registrars are required under section 41 of the Registrar of Births, Deaths and Marriages Regulations, 1987 to report unnatural deaths.

A person's death is a tragedy in itself, but unfortunately it is a part of life's cycle. People are born, they live and then they die… naturally. If someone dies before they have fully lived, this is unnatural, unless they contracted a naturally occurring disease. If, however, a person would still be alive (given the balance of probabilities), but for the contraction of a disease that they would not normally have been in contact with, or that they have had administered to them, then their death is not natural.

This raises the question: who administered the substance that caused the disease, or caused the event that put the person in contact with the disease? Below are some questions that anyone would ask if confronted with this situation. The answers to these questions will establish the circumstances around the person's death.

- Was the person in a life-threatening situation before the event?
- What disease or cause did they die from?
- Could the death be attributed to another cause?
- Are these causes naturally occurring in the population?
- Would the person have caught the disease in the natural order of events?
- Did someone's error of judgment cause the person to contract an unnatural disease?
- Was the disease administered, intentionally or unintentionally?

The inquest examines the circumstances around a person's death in depth, following strict legal processes. The cause of death is then established. As regards vCJD, the disease is considered to be the human form of BSE, an animal disease.

Not all human deaths are reported to the coroner in the normal routine of events. Many inquests are the result of a battle on the part of the victim's family. In some cases, this can take five years.

Action for Inquests is calling for consistency in inquests for those who have died from vCJD. At present, there is still doubt over whether the cause of death is natural or not.

A death from vCJD is unnatural and should be recorded as unnatural by due process of law.

- Consistency will end the battles families have in their fight for an inquest.
- Consistency will end the doubt over whether this disease is natural or unnatural.
- Consistency will ensure that the death is recorded as unnatural, with all the implications of this.

When a death is examined, due to recognition that it was unnatural, and a verdict of misadventure is recorded, it is acknowledged that the victim was killed. A natural justice has prevailed.

In finding that the victim was killed, it is understood that someone or something killed the person. Someone or something is *accountable*. An inquest is not about apportioning blame – it simply examines the circumstances. But, if those circumstances point to an unnatural death and the verdict is death by misadventure, then the blame lies with somebody.

The next stage in securing justice for our loved ones can then be pursued; namely, accountability.

Yours faithfully,
Arthur Beyless

Send your letters (or any other contributions) to: Gillian Turner, CJD Support Network, Birchwood, Heath Top, Ashley Heath, Market Drayton, Salop TF9 4QR. Email cjdnet@alzheimers.org.uk

Launch of Japanese CJD Support Network

The Japan Times reported on 26 March 2002 that the Health Ministry of Japan, the German firm B Braun Melsunger AG (which supplied human dura mater tainted with CJD) and its importer in Japan have reached a settlement with plaintiffs, who sued them for failing to ensure the safety of human dura mater transplants and for causing the onset of CJD.

Twenty CJD patients will receive a Y1.16 billion settlement and the state will pay Y3.5 million to each CJD patient who received dura mater transplants, regardless of when they had the operations. The government also said that it would prevent medical-related health disasters from happening again.

As a result of the court case, families, solicitors and researchers have worked together to launch a Japanese CJD Support Network. The network has developed a CJD telephone helpline to offer support to families who have been affected by dura mater transmitted CJD.

Gillian Turner, co-ordinator of the CJD Support Network, acted as host to Professor Kiyohiko Katahira, Junko Matsuyama and Dr Katsumi Yokosuka (George) from the general assembly of the Japan CJD Network when they recently came to Britain. They visited the Prion Clinic, the Institute of Child Health and the National CJD Surveillance Unit.

The Japanese CJD Support Network's leaflet

French CJD conference

ALMP (Association de Lutte contre les Maladies a Prions) is organising a research conference about prions and therapeutics in Paris from 1-3 December. For further details, please see http://congres.igh.cnrs.fr/Prion-Therapeutics-2002

CJD groups in America

CJD Aware

CJD Aware is dedicated to sharing information and networking. It was founded by people who had lost family members to sporadic CJD.

For further information, contact Christy C Brom, director, at info@thewayitwas.com

CJD Foundation

The president of the CJD Foundation in America, Francis Kranitz, recently visited England on a fact-finding mission and met up with the CJD Support Network. Francis lost her husband to sporadic CJD and is setting up a helpline to offer support in America.

For further information, please contact Francis at fjkranitz@aol.com

CJD Voice

CJD Voice is an Internet support group in America and can be contacted at cjdvoice@onelist.com

Concern over rise in cases of CJD in Switzerland

An article in The Lancet (Markus Giatzel et al, 13 July 2002, Vol 360) outlined how the incidence of CJD in Switzerland increased two-fold in 2001. Figures from the first quarter of 2002 indicate that the incidence continues to rise.

None of the cases fulfilled the definition of vCJD. The article stated that several scenarios could account for the increase in CJD, including improved reporting, iatrogenic transmission, and transmission of a prion zonosis.

Support agencies for the different forms of CJD

There is plenty of help available – but some agencies are more appropriate for certain forms of CJD and at different stages of referral. The chart below shows who to turn to…

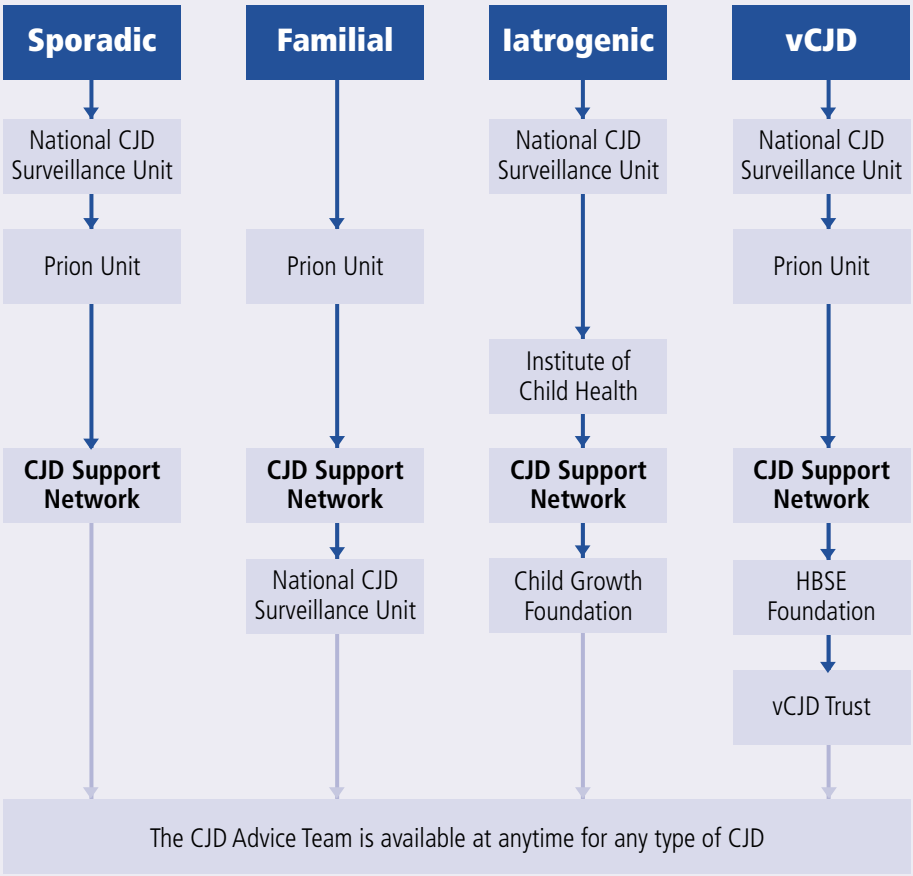
The forms of CJD

Sporadic CJD has no known cause. Symptoms include mood swings, memory lapses and social withdrawal, quickly followed by a lack of co-ordination and clumsiness. Later, blurred vision and even blindness may occur, along with rigidity in the limbs, sudden jerky movements, incontinence, difficulty in speaking and swallowing, and slurred speech.

Iatrogenic CJD is caused by contamination with tissue from an infected person, usually as a result of a medical procedure. Symptoms are similar to those of sporadic CJD, sometimes with initial depression and fatigue.

Variant CJD (vCJD) is thought to be caused by exposure to BSE in cattle. Initial symptoms are anxiety, depression, withdrawal and behavioural changes. Then persistent pain and odd sensations in the face and limbs may develop. After several weeks or months, symptoms may include unsteadiness, sudden jerky movements and progressive dementia.

Familial CJD is a very rare inherited form of the disease. The symptoms vary but are sometimes similar to those of sporadic CJD.



Contact details

National CJD Surveillance Unit
Care Office
Western General Hospital
Crewe Road, Edinburgh EH4 2XU
Telephone Number: 0131 537 3073

Prion Unit
St Mary's Hospital
Praed Street, London
0207 886 6883

Institute of Child Health
39 Guildford Street
London WC1N 1EH
Contact: Leah Davidson
020 7242 9789

Human BSE Foundation
Chairman: Lester Firkins
Secretary: Frances Hall
Helpline 0191 389 4157

CJD Support Network
Contact: Gillian Turner, Network co-ordinator
Helpline 01630 673993.

CJD Advice Team
Contact: Care Office,
National CJD Surveillance Unit
(as above).

vCJD Trust
Charles Russell, Solicitors
Contact: Edwina Rawson
0207 203 5335

How our daughter Jo was supported

By Janet

It has saddened us to hear of the difficulties that many CJD patients have in obtaining the care and support they need. While nothing can mitigate the devastation of our daughter Jo's illness, we have at least been blessed with caring support on all sides. We'd like to offer this encouragement to other patients and their families: whatever support you need can and must be made quickly available. If you have difficulties or delays, call one of the national CJD care co-ordinators at the National CJD Surveillance Unit; they're there to make sure that your local agencies know what is needed, to stir them up if they're slow to respond, and, if necessary, to provide funding from the national care fund.

We're also happy to discuss our experience with you if it will help. While we do not yet wish to be publicly identified, inquiries through the CJD Support Network or the Human BSE Foundation referring to this article will be passed on to us.

Early stages

There were delays in getting appointments in the early stages, and diagnosis was slow in coming; but not beyond the targets set for health services in general. Our biggest disappointment was with the education psychology service, which had a waiting list of 16 weeks when Jo's behavioural disturbance was reaching dangerous proportions. This forced us to rely on the medical rather than the psychological route (only eight weeks to be seen by a consultant paediatrician!) In retrospect, this was probably an advantage, in that there was no initial psychological or psychiatric diagnosis to delay recognition of vCJD. I think we were also lucky in that, when we finally did see our consultant, Jo was in transition from behavioural to physical symptoms.

Since we have had a diagnosis, everyone has pulled out all the stops for Jo. Professionals have all been willing to commit the level of support she needs, and fear of infection has not undermined the quality of her medical and personal care. Everyone has been keen to learn as much as possible about the condition, both for Jo and for any future cases they may encounter.

Medical support

Our local hospital has taken advice from CJD professionals at St Mary's Prion Clinic and the national CJD care co-ordinators, and has worked closely with them and with us to give Jo the best care possible. Nursing, medical and teaching staff have welcomed Jo as a member of the 'family' on the children's ward – on our weekly day visits, staff not involved in her care will stop to speak to her, just because they care. And this is from a hospital currently in the news as a 'failing NHS trust'!

When trials of quinacrine became available, we chose to take part in a formal trial through St Mary's Prion Clinic, mainly because of the close monitoring that would be provided. This proved valuable when blood tests indicated possible liver problems as a side-effect – close monitoring of the disease showed no sign of improvement, so the decision to stop treatment was a relatively easy one.

The children's ward and the Prion Clinic at St Mary's have given us very good service. There have been some managerial problems, some of which were due to the ward staff's inexperience with vCJD (Jo was St Mary's first child case); others reflect more general problems in ward management, including heavy dependence on agency nurses. However, almost all the medical, teaching, nursing and support staff are very supportive, both to Jo and to us, her family. The Prion Clinic has been wonderful, keeping us well informed and producing a small miracle in getting the quinacrine trials formally set up in record time, even though the delay seemed interminable to us.

The only major failure in medical care has been from our GP, who had no idea what to suggest when Jo's behavioural problems first arose, and who has shown no interest in her case since the initial referral, not even when the diagnosis was given to him. Fortunately, the excellent hospital care has meant that we have had little need for GP support. We have recently changed to a new practice, and we now have weekly visits from a new GP (alternating with visits to the hospital).

Community support

Full marks also go to our local children's social services – I sometimes think Jo must be the best looked-after girl in the country! Our key worker has kept everything running smoothly for us. From the start, social services have liaised with the hospital ward, Jo's school, the education service and us; and they have taken advice from

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the Prion Clinic and the national CJD care co-ordinators. They have planned ahead for Jo's likely needs, letting us know what support we may require, but always leaving the final decision to us.

When Jo was still fairly active, she had access to the social services' excellent special needs day centre, recently praised in the government's white paper, *Valuing people*. Initially, they provided respite care for a few hours a week, either through home sitting or visits to the centre to enjoy their sensory room, soft play facilities and music centre. She also had swimming (as long as she was able) and hydrotherapy.

As Jo's needs increased, we opted to have care workers in the home. We began with three nights a week, and built this up gradually, as required. We now have a team of nine carers providing cover 21 hours a day, 7 days a week. This includes two carers during the day for 'moving and handling', so that Jo's care is not restricted if her Dad and I need to be out of the house. Every one of the carers is totally committed to Jo – we had no break in cover over Christmas, for example. Between them, they offer a range of 'extras', from massage and aromatherapy to French plaiting and nail painting; and, with their varied interests and personalities, they provide a variety of company that we, with all the love in the world, could not. Our key worker also makes sure that we have the equipment Jo needs in good time. A special wheelchair and hospital bed were ordered well in advance, so that they were quickly available as soon as she needed them. A fully adjustable armchair, initially not at all successful, has now come into its own and gives Jo regular breaks from her bed. We did have some trouble with the hoist, which was thrust upon us long before either the carers or we were ready to give up lifting Jo – but that too is now, sadly, indispensable. Our specially adapted vehicle took a while to obtain because our key worker didn't know that funding could be obtained from the National CJD Care Team in Edinburgh for that; but once we'd passed on information from another patient, she got it organised for us. We no longer have to wait for overcrowded ambulance transfers to and from hospital, and we can take Jo for drives in the country whenever she's up to it.

Alternative therapy

We were advised early on to find aromatherapy for Jo, as this has benefited patients in the past. Luckily, our first daytime carer could offer this, along with massage and 'healing', so Jo can have different treatments as and when needed, without having to visit a specialist. If you do use a specialist, funding can be provided either locally or through the national care fund.

The combination of massage and aromatherapy has been hugely important for Jo. Throughout her illness, they have improved her general comfort and physical well being; the constant 'pampering' has also been invaluable in keeping her spirits up. The excellent condition of her skin, and the constant attention she receives, are also the best possible protection against pressure sores.

Support for the family

Finally, don't forget your own needs. Jo's medical team has no direct responsibility for us, but has always been there if we need to talk. If we need more specific support (for example, counselling), they're able to advise us on where to find this. Jo's key worker can also facilitate access to such services, as well as more practical matters such as benefits or home help. They even sent in a team to help us clear our downstairs room so that Jo could sleep there when it became necessary. If we needed respite, or could no longer care for Jo at home, members of the team would be there, ready to make the necessary arrangements.

Don't be afraid to ask

Of course, not everyone will need or want the same support that we have chosen. A good service will let you know in advance what needs might arise, and what services or facilities are available to deal with these. They should then leave you to choose the help that you need. Don't be afraid to ask about anything that's not offered, however unusual it may seem – our key worker has even helped Jo's best friend to get support through the education service because she was so disturbed at seeing her pal deteriorate so quickly.

Potter magic

Jo discovered Harry Potter when in the early stages of her illness, and read all four books twice before reading became a problem. We had, therefore, very much looked forward to the film. In November, realising that the film had arrived too late for Jo to go out to the cinema, our friend Karen had the brilliant idea of trying to get an advance video.

The Make a Wish Foundation made enquiries for us, but initially things didn't look too promising. Then the Foundation contacted the Warner Brothers' production team, which was working on the next film, to ask whether Jo could visit the set. Their response was

immediate and overwhelming. A senior manager stayed late one night to make a video copy for Jo; three managers brought it round, with a large screen TV to make sure she'd get a good view. They couldn't leave the video with us but they were happy to leave the equipment and bring back the video as often as Jo wanted it. Jo amazed us with her staying power, watching the whole film in two sittings, and then again twice at one sitting each. Warner also brought Jo some amazing goodies, including an autographed poster, crew T-shirts and a cast sweatshirt. Just one more example of what can happen when hearts are touched.

When we applied to the Make a Wish Foundation, we had to give three wishes in case our first couldn't be granted. Our second wish was for some sensory equipment of the kind she had access to in our local children's special needs centre. They sent her a few items even though the Harry Potter wish had been granted – most effective was the rotating disco light that we regularly use, especially at bedtime.

Our third wish was to obtain in UK format a video of a US band show called Blast. Jo was a member of our local youth marching band, and she'd gone with her band to see this spectacular show the previous year. We ordered the US video over the Internet, and made enquiries about getting it converted. While we were still trying to find out where to get copyright permission for the conversion, one of Jo's carers tried putting the tape in our video to see what happened, and it worked perfectly! So, how's that for three wishes granted?

A difficult diagnosis

CJD is rare and health professionals do not always recognise it. A trustee and branch member of the Alzheimer's Society writes of the experiences of one south Gloucestershire family.

Janet Withall

CJD affects only one person in a million in the UK. Only 50 new cases are reported every year, but because of its symptoms, even the professionals often confuse it with dementia.

In June 2001, Jill's family first noticed that something about Jill was changing. She lost her temper more quickly than normal, and quickly changed from being nice to nasty. In August, she started getting lost when driving. She began to call her sister, Dawn, and her sister-in-law, Brenda, to tell them about lost keys, lost glasses and even lost teeth, which were usually found in the most obvious, or ridiculous, places.

Jill began to miss meals, not get dressed and not keep her flat clean. Her family were becoming increasingly concerned, and when Dawn received a call from Jill's local shop describing her sister's odd and erratic behaviour, she rang the GP and took her in to the surgery. Jill was referred to a specialist and given an initial diagnosis of dementia, with a recommendation that further tests such as an MRI scan, a CT scan, a lumbar puncture and blood tests were carried out.

Jill returned home but her condition continued to deteriorate. When Dawn called to take her sister to church and found her waiting in her underwear, she rang the doctor again and he had Jill admitted to Frenchay hospital. During her stay in hospital, Jill had all the tests that had been requested by her specialist except the MRI scan, but the results all came back negative. She wandered from ward to ward and was unable to find her way back to her bed from the toilet. After a week in hospital, Jill was discharged, although she lived alone, and was sent home with vitamin B12 supplements.

'We got the feeling that the specialist was beginning to think that Jill's problems were drink related, whereas we knew that she hadn't even touched alcohol for months. The problems continued: burnt food, inappropriate meal times, losing things – but when we talked to her about these things she always had an answer and was very good at covering up her mistakes. She successfully managed to hide all her problems from her friends; they were amazed when they finally found out the full extent of her confusion', explained Dawn.

When Jill's behaviour deteriorated to the point where she was trying to get into other people's flats, was locking herself out, and was wandering around outside at night, she went to stay with Dawn and her husband, Martin.

'While she was staying with us, I found her awake at 3am, so I offered to make her a cup of tea. When I went back upstairs, I found she had got into bed with my husband! He was somewhat shocked when I walked into the room with two cups of tea while he was snuggled up with someone he had clearly assumed was me,' said Dawn. CONT

Carers' stories

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When Dawn's daughter invited Dawn and her husband to visit her in the USA over Christmas 2001, the family had to look for respite care.

'Because Jill would be self-funding, social services did not have to get involved in helping us but we eventually did find a place in Somerset. It was a very nice home and the staff were lovely but it was difficult to leave Jill, who just wanted to come home. Jill was now beginning to be very unsteady and had a few falls at the home; she was also becoming incontinent and Martin, Dawn's husband (who had been a farmer), likened Jill's symptoms to those he'd seen in cows with BSE,' explained Jill's sister-in-law, Brenda.

Dawn's daughter did some research on the Internet and found the number of the National CJD Surveillance Unit at the University of Edinburgh. Dawn called them and they sent her the relevant information.

'When I read the information they sent from Edinburgh, I just thought that Jill's symptoms fitted precisely with a diagnosis of CJD – I mentioned it to Jill's doctor and he just said that I was making it fit, but I was convinced'.

Jill was admitted to Frenchay for tests, which duplicated the tests already done – again they revealed nothing. By now, Jill was doubly incontinent and could barely move. She was too much for the residential home to deal with and so was again taken to Frenchay hospital.

When Dawn met Jill's specialist at the hospital, she mentioned her thoughts about CJD. The specialist commented that she had 'done her homework' and got in touch with the National CJD Surveillance Unit in Edinburgh. He also did some more tests, including a special lumbar puncture, and sent a sample of cerebrospinal fluid to Edinburgh. Shortly afterwards, the family met with Jill's doctors who confirmed that they were 95 per cent sure that Jill was suffering from sporadic CJD (a definite diagnosis cannot be made until after death). Since Jill no longer required hospital treatment, but was not yet not in such an advanced state that Macmillan nursing was required, Jill's family were again in the difficult position of finding somewhere where she could be looked after.

'It was very difficult to find somewhere and actually in the end a social worker found her a place at a nursing home in Kingswood. The matron of the home came to see Jill in hospital and it was only then, when talking to a nurse, that I discovered that the hospital had seen other cases of CJD, which did make me angry about the difficulty we had had in getting Jill diagnosed'.

Jill moved to the home in Kingswood, where someone from the family visited every day. She developed difficulty in swallowing; she had no leg movement, and she could not speak anymore.

'The whole process has been a complete nightmare. At least we have a group of family members to help deal with the whole thing, but frankly it has taken over our lives for the last year,' concluded Dawn.

Jill finally died on 5 August at the age of 66. Our condolences go to the family, and Janet would like to thank Jill's family for their co-operation over this article.

CJD Support Network membership application

Becoming a member of the CJD Support Network adds to our strength and enables you to take a full part in the decision-making process and the work of the Network.

☐ I would like to become a member and receive the CJD Support Network Newsletter.
☐ I would like to become a member but **not** receive the CJD Support Network Newsletter.

There is no fixed subscription, but please give generously to help our work.

☐£8 ☐£12 ☐£25 ☐£50 ☐Other .....

Please make cheques payable to **CJD Support Network**

However, if you are a carer and would appreciate free membership, please tick the box ☐

Name Title .....

Address .....

.....

Postcode Telephone .....

I am caring for someone with CJD: ☐ at home ☐ in residential care

I am: ☐ a concerned relative/friend ☐ former carer ☐ professional ☐ interested NOV 02

CJD SUPPORT NETWORK

The views expressed in this Newsletter are personal and not necessarily those of the CJD Support Network.

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