

The CJD Support Network is 10 years old

The CJD Support Network is 10 years old. It is an important milestone for us as we celebrate ten years of providing practical and emotional support for families and professionals affected by CJD.

With your help the CJD Support Network is now recognized as the leading charity supporting families and professionals affected by all

strains of Creutzfeldt-Jakob disease. 2006 also heralds the start of an important decade in CJD, which has seen new challenges, knowledge and hope for a future treatment for this devastating disease.

With the new concerns and notifications to donors surrounding secondary transmission through blood and surgical instruments, we feel it

is timely to arrange a conference 'Decade past, decade to come'. It is our plan to bring together families and professionals affected by all strains of CJD and for those new people who have been informed that they are at a higher risk of CJD through secondary transmission.

The conference, on Monday, 20 March 2006, at BMA House in London, will also mark the 10th anniversary of the announcement in the House of Commons, by Mr Stephen Dorrell, the then Secretary of State for Health that ten young people had contracted a new variant of the harrowing, and invariably fatal, CJD.

At the conference, to mark our 10th year we will also be commissioning a piece of research with Lancaster University to look at the information, support and care needs of patients and families affected by sporadic CJD.

If you would like further details of our exciting plans for 2006, please contact the Network either by telephone on 01630673993 or by email info@cjdsupport.net.

Blood transfusion risks assessment

Source: 20 July 2005 press and Department of Health statements

In July 2005 around 100 blood donors were told that they may have a greater chance of carrying the vCJD agent, compared with the general population as their blood had been donated to three people who had since died of vCJD.

Although it is not known whether the source of vCJD in the patients who have died is related to the blood that they received, precautionary steps were being taken to inform and provide support to the 100 individuals who had donated blood as well as safeguard public health. This is being done on the advice of two expert committees and a detailed risk assessment exercise.

As a precautionary measure the donors were told not to donate blood, tissue or organs and to inform health professionals so extra precautions could be taken should they themselves have surgery or other invasive procedures.

The Chief Medical Officer, Sir Liam Donaldson, said: 'We need to ensure that appropriate action is taken on any new information that becomes available on the risk of transmission of vCJD, to protect the public as much as possible. When a recipient of a blood transfusion goes on to develop vCJD, we have to consider the possibility that the infection could have been passed on through the transfusion. Until a reliable blood screening test becomes available, it is sensible to proceed with highly precautionary measures such as this to rule out any possibility of onward transmission of the disease. We are committed to further research to help understand this disease and diagnose infection at an early stage.'

'Following the identification of vCJD, we introduced a number of measures to reduce the possible risk that infection could be transmitted through the blood supply. Since the announcement in December 2003 of the first case of possible transfusion-associated transmission of vCJD, we have further strengthened these preventative measures. The decisions taken so far have been based on the principles of caution and openness. This announcement is a continuation of the process.'

Dr Angela Robinson from the National Blood Service said: 'Blood donors are highly committed to helping other people and we greatly value their

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contribution. The NHS depends upon their continued commitment in order to be able to save lives. This notification exercise will affect in the order of 100 donors. If you have donated blood in the last five years and are not contacted shortly, you can be assured that you are not involved in this new safety measure and need to take no further action.

'For those people who are involved, this information may be difficult to absorb. That is why we have set up the National Blood Service helpline and are working with their doctors and other clinicians, to ensure that they have the information and support they need.'

See also 'Blood donors who gave to CJD cases' on page 15

International CJD Day

To mark International CJD Day on 12 November, the Network held a highly successful family support meeting and AGM at the Glaxo Neurological Centre in Liverpool.

Families affected by vCJD, sporadic and familial CJD came together to share experiences and insight into the symptoms they had witnessed and to give support to current nursing families.

Our chairman, Dr Angus Kennedy, taught attendees how to be a neurologist by giving an insight into how a neurologist reaches a diagnosis. He said listening and observing the symptoms displayed gives a clue as which part of the brain is most affected.

vCJD in Europe

Holland

On 3 May 2005 Reuters reported the death of a 26 year old woman through vCJD in Holland. This was the first case of vCJD in Holland

and was first diagnosed in April by specialists at the Erasmus Medical Centre in Rotterdam.

Portugal

First probable case of vCJD has been reported in Portugal and has been confirmed by the UK National CJD Surveillance Unit. The patient, a 12 year old boy, does not have a history of travel to the UK.

Ireland

Eurosurveillance reported a further probable case of vCJD in Ireland. Three cases of vCJD have now been reported, although the first in 1999 was thought not to be indigenous as the patient had lived in England for several years. Eurosurveillance also reported that a blood donor was being treated for suspected vCJD in a Dublin hospital.

Spain

A first case of vCJD has been reported in Spain, according to Eurosurveillance's weekly report. The Spanish Ministry of Health reported that the patient, a 26-year-old woman in Madrid died in July 2005.

CJD Support Network commissions research into sporadic CJD

To recognise ten years of providing practical and emotional support for CJD, the CJD Support Network is commissioning a piece of research in conjunction with Lancaster University. The research will focus on the information, support and care needs of families affected by sporadic CJD.

The research will be launched at the International Conference 'Decade past, decade to come' on Monday, March 20 2006. The research findings will be announced at the CJD Support Network's AGM in November 2006.

If you would like to take part in this research, please contact Gillian Turner on 01630673993.

The Network is grateful to Stuart Durkin and friends who have agreed that the money raised during 'Wheelin for Willie' will be used to sponsor this research. Stuart's father, Willie, died of sporadic CJD in August 2003.

New leaflet

As part of our ten year review we are producing a new leaflet on the work of the CJD Support Network.

We are now recognized as the leading charity for all forms of CJD and as our work is expanding to support patients who are told they are at a higher risk of CJD through secondary transmission, it is timely to revamp the charity's leaflet.

We are very grateful to Florence Tittensor and her friends who have raised the money, in memory of Florence's husband Alan, to fund the new leaflet.

Recent CJD figures

The number of deaths of definite and probable cases in the UK, up to 2 December 2005, from the CJD Surveillance Unit in Edinburgh.

Year	Sporadic	Iatrogenic	Familial	GSS	vCJD	Total*
2003	77	5	4	2	18	106
2004	52	2	3	1	9	67
2005	55	3	4	4	5	71

*As at 2 December 2005



Name that neurologist

Sarah Ridgewell has knitted a fabulous clown, in the image of a doctor. Our Christmas fundraiser is 'name that neurologist'. For a small donation, suggest a name for the doctor. You never know, you may win this fabulous knitted clown for a loved little one. So far, we have had some 'unique, thought-inspiring' suggestions! But you just may have the winning name. The winner will be notified in the new year.

Send your suggestions to
Gillian Turner
CJD Support Network
PO Box 346
Market Drayton
TF9 4WN

Wheelin' for Willie

Stuart Durkin lost his father Willie to sporadic CJD on the 30th August 2003. Stuart now sits on the CJD Network management committee and at the end of May 2005, embarked upon a sponsored bike ride through which he raised funds for the Network.

The bike ride followed the historical pilgrim route the Camino de Santiago, from Roncesvalles in the Pyrenees to the cathedral city of Santiago in Galicia, some 850 km to the west. Stuart and his friends completed the route within seven days.

This route is of symbolic significance to Stuart. Prior to his father's illness Stuart had lived in the Basque city of Bilbao for three years, after meeting a Basque girl during a post-graduate degree at Leicester University. Tragically, the Basque girl, Izaskun, lost her mother to the genetic form of CJD a matter of months before Stuart's father died.

The route passed close to the Basque country and Stuart lit a candle at the Cathedral in Santiago for his father, Izaskun's mother and all victims of CJD.

At the end of the ride: Stuart Durkin with Phil Goodwin and Eddie Kynes in front of the cathedral in Santiago



Flora Walk for Life

Anne and Louise

On Sunday 5 September 2004, we took part in the Flora Walk for Life 5 km run in Hyde Park. We decided to take part to raise money and awareness for the Human BSE Foundation and the CJD Support Network. Both of these organisations rely on charitable funding to survive. After the loss to CJD of a close family member, Andrew Keenan, our family benefitted from the continuous support offered by these organisations – as have many other families of the victims of this terrible disease.

The race was held on the first anniversary of Andrew's funeral. The day felt more poignant because of this. Out of the thousands of competitors, we were the only two supporting our charities. We could not believe the number of people who had not heard of us, we also had quite a number of people approach us. The race itself was an incredibly emotional journey for both of us; we were very aware of the reason why we were doing this.

To complete the race was a huge sense of personal achievement on every level. We cannot explain the feeling of crossing the finish line with the noise of the cheering and clapping in the background, it is something that will stay with us forever.

Andrew qualified as a chef and worked in a number of top restaurants throughout the country. He enlisted into the Army as a chef, and was promoted to Lance Corporal. He was a lively young man with a fantastic sense of humor, who was always up for any social occasion. Andrew was loyal and deeply passionate about the important things in his life. He would always achieve anything he put his mind too, he had such sheer determination.

Andrew was diagnosed with vCJD on the 1 July 2003 at the age of 25. He passed away on the 25 August 2003, aged 26. He is forever in our hearts.

We would like to thank our families and friends for all their support and to everyone who sponsored us. We raised £660, which we shared equally between the two charities.

Dear editor

My mother died from CJD in June 2004. In the year or so since I have had time to reflect and can tell you that I was very grateful for all the information available from the CJD Support Network at the time of the diagnosis. Also, the support that was provided by the helpline was critically important for our family and critically improved the care we were able to provide her.

It seems to me that there are many ways in which help and support for the victim and their carer could be improved. Elsewhere in this edition of the newsletter I outline the events that have led me to this belief. (See my article 'Make it go away' on page 8.)

As we cared for my mother, we learned how to improve her comfort by default. For instance, we found that as her eyesight deteriorated that her confusion was increasing. If we wore clothes with patterns she could not quite interpret the pattern and sometimes these frightened her. So we vowed to wear plain clothing. Also, we found that it was best to crush her tablets and sprinkle them in lemonade – easier to swallow and probably better tasting. At night we found that switching off her bedside light only served to alarm her if she awoke during the night. By leaving it on she could more easily establish what was going on around her.

No one knows what happens to the senses of the person who is suffering from CJD and there is the possibility that each person's experience of the disease is unique. However, it seems that nothing is lost by offering such common sense advice to those who are newly plunged into the darkness of caring for a person with the disease.

Through the CJD Support Network, I would like to

Thank you all!

The CJD Support Network is very grateful for all the public donations it receives and every penny is directly used in our work to help and support families affected by all strains of CJD. Below is a sample of donations raised by members of the network in 2005. You will see that these amount to more than £8,500, which represent a great deal of dedicated hard work. Thank you to everyone who has donated.

Stuart Durkin – 'Wheelin' for Willie'	£2,850.70
Victoria Pennell – London Marathon	£1,371.49
Sale of cakes by Maria	£39.00
Fund raising party, Tracy	£600.00
Smarties appeal	£439.00
Tracy Huggett's social evening	£387.04
Anne and Louise O'Connor – Flora Walk for Life	£330.00
Florence Tittensor – coffee morning	£640.00
Liz Burgess – bowling fun day	£1,007.45
Pauline Taylor – sponsored walk	£700.00
Lynn Walton – Youth Justice Board Roman walk	£365.00

compile a list of similar advice based on the experiences of carers. This could be put together in a booklet to provide basic guidance that might ultimately improve the comfort of those in the terminal stages of the disease.

If you have such experience and would like to share it, please contact me through Gillian Turner of the 'Network' (or email me directly at: bradley449@yahoo.com). Any contributions can be in confidence or in memoriam. Until that bright day dawns when a cure is found, then the best that can be offered is the comfort of the patient.

Yours faithfully,
Mike Bradley

Dear Sir

Thank you very much indeed for the caring grant of £200 which you kindly sent me. It was indeed of great help towards the extra costs of my late wife's funeral – in fact, I could not have managed without it. Thank you again for your care and your support.

Yours sincerely,
(name withheld by request)

Dear Sir

I am writing to thank you for the cheque you sent to me. It will certainly help with costs. I am still in terrible shock at having my husband taken from me by this terrible disease.

Yours sincerely, (name withheld by request)

Dear Gillian

Thank you for making us feel so welcome and at ease at Saturday's meeting. We found it extremely helpful to listen as to how other families had coped and it gave us strength to face the future which we realize will be very difficult indeed. All being well I shall try to attend the BMA meeting and look forward to seeing you there.

Yours sincerely,
(name withheld by request)

Send letters to:

Gillian Turner
CJD Support Network
PO Box 346
Market Drayton
TF9 4WN

Caring for Dad: theory and reality

By Angela

In the summer of 2001 we were finally able to put a name to Dad's illness; GSS or familial CJD. That was the first challenge. The second was to look after him and that by far was the greatest challenge of all.

The timing and quality of advice we received from most individuals and organisations we encountered was good. However, what we found in practice was that the timing and quality of practical local support was extremely variable. Armed with facts and knowledge about what should be available, our resolve to seek that support was strengthened. Thank goodness Dad had loving family members around him to give him a voice and forward his best interests.

Financial support from the State benefits system was easy to identify but we found the form-filling daunting and very time consuming.

Every few weeks the family would meet up with a team of

experts to co-ordinate the type and level of support needed as Dad's illness progressed. This 'case conferencing' was good for two reasons: firstly, we could try to anticipate needs and secondly, it gave Mum, who was the primary carer a sense that we were not alone.

Nonetheless, despite the discussions and plans, in practise local support was often slow to materialise.

Social services agreed carers in the home were needed. This service was out-sourced to an agency which didn't have enough workers. I had to threaten social services with a formal complaint before they started to use their initiative and employ another agency.

The local authority approved the grant funding for a ground floor bathroom but weeks passed before the tenders were processed. Alas the builder that was selected proved to be a 'cowboy' and walked out on the

job. Finding a new contractor was time consuming and upsetting. We really could have done with the local authority being far more involved and assertive at every stage of the build; in the end the bathroom was completed two weeks before Dad died.

We are eternally grateful to a number of key professionals. Everyone strived to give Dad 'quality of life'. On some occasions, bureaucracy, administration, lack of finance and poor communications presented obstacles. Then, when we were already consumed and exhausted from looking after Dad we had to find some more fight in us.

Dad passed away peacefully three and half years after his diagnosis. There was always one of us with him night and day to keep him out of danger; to safeguard his quality of life and dignity. In that respect he was a very lucky man. But what happens to others who do not have that network of family support?

Nursing a vCJD patient at home

By Claire McGee

The first knowledge of my patient was when I was invited to a case conference by the GP for a patient with a neurodegenerative condition. The importance of attending was emphasized. This case conference started with a presentation from the CJD surveillance unit co-ordinator. At this point I didn't know what to think. I had heard about vCJD via the media like most people. Now I faced the challenge of nursing this young patient.

I met the parents and they were definite in their views of how their child should be cared for. Only physiotherapy, occupational therapy and social services were required at this stage. The parents wished to carry out all the nursing care themselves. I had limited input to give advice regarding urinary incontinence and skin care, and it was then that I met the patient.

Following a chest infection approximately three months later my patient's condition deteriorated. After a period of hospitalization, they were transferred home. The patient, who was unable to communicate, was totally dependant on others for all activities of daily living. All professionals involved felt that the terminal phase of the illness had begun. The appropriate equipment was organised for the home and the family trained in caring for the PEG tube [a feeding tube placed directly into the stomach], the feeding regime, skin care and suctioning.

At this point infection control became a major issue. Naturally there was a lot of fear from staff, both hospital and community, regarding the possible transference of the prion protein. Through liaison with the CJD surveillance unit, infection control specialists and the family, the appropriate measures were put in place – measures to protect staff, patient and family members. At times it was difficult accessing the correct information as research was, and is, still ongoing, with different specialists within the nursing and medical community having different ideas on appropriate measures – as had the family. Through my own research and

accessing documentation from both the CJD surveillance unit and the Department of Health, measures were implemented which minimised the risk to all concerned, but prevented the patient being isolated and losing a sense of normality. An education programme was undertaken with staff members to ensure infection control procedures were clear. This also enabled me to discuss any concerns the staff had.

Even though it was felt that the patient was in the terminal phase of the illness the parents were not prepared to give up. In their continual search for treatments they required a lot of support – physical, psychological and emotional – as they also had the rest of their young family to care for. At times, visits to the home involved looking after every family member and a lot of uncharted territory was covered. This proved to be a large and quick learning curve for myself as, at times, I was the district nurse, social worker and counsellor.

A package of care was initiated to assist the family as the patient's needs were great and the family was becoming exhausted. 24-hour care was now needed. Due to the frequent choking episodes which required suctioning, it was decided that qualified nursing staff were needed. The package was led, and is still maintained, by the district nursing service. As this required a lot of time to organise, it was at this point that my manager became involved in the day-to-day aspects of care. She also gave me support. Caring for the patient was quite intensive as I had been the only district nurse involved. At times it was stressful because of all the new challenges.

Due to the intensive nature of the care required, additional district nursing staff were soon introduced, with the family's permission. Now a nurse is delegated to manage the package of care on a day-to-day basis. This enables me to provide the appropriate level of care to the other patients on my caseload. To ensure all staff involved are carrying out the

same level of care, regular training is organised by district nursing in conjunction with the occupational therapist and physiotherapist.

This level of care continues today and for the foreseeable future as my patient is considered no longer to be in the terminal phase of vCJD. The challenge now is continuing with the high level of care which is needed to prevent problems occurring with the PEG tube and feed, skin breakdown, constipation, urinary incontinence and carer breakdown. Independent counselling sessions have been organised by my manager, due to the intense nature of the situation. These help the staff to manage their own concerns, thoughts and feelings.

Nursing a patient with vCJD is an interesting and challenging experience. When I first became involved I was venturing into the unknown. That continues today as more is discovered about vCJD. This experience I am able to share with the other health professionals involved. Most district nurses have never been in contact or nursed a patient with this condition and therefore it is a huge educational experience for the whole team. My role has expanded to become an educator of CJD for other district nurses and nursing students. I am fortunate to have good support systems, namely my manager and the CJD surveillance unit co-ordinator, as without this the challenge would be much more difficult and stressful.

Claire McGee

District Nursing Sister, North and West Belfast Health and Social Services Trust

EDITOR'S NOTE: In 2002 the CJD Support Network commissioned The Queen's Nursing Institute to write 'INFORMATION AND NURSING CARE GUIDELINES FOR CJD'. A copy of these guidelines can be obtained from the CJD Support Network at a cost of £5. Free to family carers.

Fatal familial insomnia

An overview of this rare prion disease

Chris Rhymes, Clinical Nurse Specialist at the National Prion Unit

There is a move in the scientific community away from using symptomatic descriptions of inherited prion illnesses (such as Gerstmann Straussler Scheinker, fatal familial insomnia and familial Creutzfeldt-Jakob). This is because the same mutation can present with different symptoms and it is more accurate to call the underlying diseases after the mutation itself (eg P102L, D178N, E200K). However fatal familial insomnia is most commonly associated with the D178N mutation.

Characteristics

Fatal familial insomnia is a progressive and fatal form of prion disease, characterized by disturbances in sleep, autonomic function and progressive cognitive decline.

Fatal familial insomnia (FFI) was first described in 1986 by Lugaresi, in his description he tells of a family physician who was investigating a family member with a 'peculiar, fatal disorder of the sleep'.

FFI is rare, for example from 1986 to 2003 there were only 60 cases reported world wide, and the countries included Italy, United States, Germany, Australia, Japan, Canada and the UK.

Onset of the disease typically occurs between the ages of 40-60 years.

Gene mutation

FFI is an autosomal dominant disorder, specifically a mutation at codon 178 of the prion protein gene (D178N). In FFI, a mutation at codon 178 of the prion protein gene (PRNP) results in the substitution of the amino acid, asparagine, for aspartic acid. If the normal allele is methionine then the phenotype is usually FFI. If it is valine on the

normal allele then classically it presents as familial CJD.

However, a similar non-familial phenotype can occur spontaneously, Gambretti et al (1993), reported a patient with FFI type symptoms but without mutations in the PRNP, and described this as a case of sporadic fatal insomnia.

Symptoms

Neuropathologically FFI is also different from the other prion diseases. Whereas CJD attacks primarily the cerebral cortex, FFI attacks the thalamus, specifically the anterior and medial thalamic nuclei. The thalamus acts as a relay station, co-ordinating the transfer of information between the cerebral cortex and lower brain pathways. During sleep it is thought that the thalamus modulates cerebral activity allowing for the vegetative state of sleep to come over an individual.

One of the first symptoms of FFI is an altered sleeping pattern which progresses to insomnia. This in turn gives rise to loss of vigilance, progressive dream states, hallucinations and fatigue. In the initial stages of the disease intelligence is often intact, so patients are aware of symptom progression. As the disease progresses the sufferer shows signs of memory impairment, deficits of attention, depression and progressive dementia.

FFI also causes disturbances in autonomic function (dysautonomia) this may manifest as elevated blood pressure, episodic hyperventilation, excessive lacrimation, sexual and urinary dysfunction, and/or a change in basal body temperature. Weight loss with no decrease in appetite is also a reported symptom.

In some individuals signs of brainstem involvement may become present. These include eye movement difficulties, dysarthric speech and ataxia affecting mobility. Pyramidal and extra pyramidal signs are common and myoclonus may also be present in the later stages.

Diagnosis

Investigation for FFI includes polysomnography which shows loss of delta activity during sleep, loss of REM phase and progressive flattening and slowing during wakefulness. MRI often appears normal, though PET scans show reduced uptake in the thalamus. Genetic testing confirms clinical diagnosis.

Treatments

There is no effective treatment for FFI, although a study is underway to examine the efficacy of drug therapy (the MRC Prion-1 clinical trial at the National Prion Clinic). Pentosan polysulphate is being used in some individuals with prion disease. Symptomatic control is of paramount importance to help alleviate the suffering of the individual and their families.

Genetic counselling and testing is available for family members. This will provide individuals and families with information on the nature, inheritance and implications of the condition to allow them to make informed medical and personal decisions. Presently testing is only available for adults over 18 years of age.

Any other inquires about FFI or any other prion diseases please email the National Prion Clinic at prion@uclh.org.

'Make it go away'

**A heartfelt account by Michael Bradley
of the progress of his mother's sporadic CJD**

It has been just over a year since my mother died from sporadic CJD in June 2004. This has been an intense period of difficult reflection as I try to fathom what happened and why it happened. The first signs of the disease had only surfaced in November 2003 when Mum began to forget a word or two mid-sentence. Nothing too peculiar about that, I forget my words all the time. Mum and the family just put it down to age – she was 60. By early 2004 more symptoms were appearing. A good car driver all her adult life, Mum began to have difficulty in judging the distance of oncoming cars and so she stopped driving. Then she started to lose weight and had bouts of nausea. More forgetfulness followed and she was referred to a neurologist and on to a psychiatrist who could find no obvious condition that could be causing these problems. By April, Mum was having some difficulty walking. A return visit to her GP resulted in the frighteningly ironic diagnosis of 'It's all in the mind'. So near, and yet so far – it wasn't in the mind it was in the brain.

May 2004 and in sheer frustration my stepfather changes Mum's GP to one at another surgery in the small Welsh village in which they lived. This second GP agrees that whatever is happening needs more investigation and an appointment is made at an outpatient's clinic at the local regional hospital. However, the rate of deterioration was becoming so rapid that Mum was admitted to hospital as an in-patient on Friday 14 May. She was able to walk onto the ward and in that first weekend she even helped some of the older patients whose mobility was worse than her own. She always offered a helping hand to strangers.

All sorts of tests were being done, including an MRI scan. All of these were throwing up negative results, yet all the time she continued to decline. Within those first two weeks in hospital Mum lost her mobility and the ability to swallow solid food and could no longer feed herself. Then the tremors started. With all testing options beginning to dry up, an EEG (brain) scan was performed on Wednesday 26 May and the cause of the problem was immediately apparent. On Friday 28 May, my stepfather was called into an office by the specialist and was told that his wife had CJD – an acronym for a disease with a long, almost unpronounceable foreign name. At that time, no one in the family knew much about this disease. Now, we are lay experts in something that, to be frank, we had no desire to be experts in.

No-one in the family knew much about this disease

Stepfather was in a daze, but was assured that the family would receive help and support from a specialist unit in Edinburgh. It was bank holiday weekend so we all gathered around in the hospital not really knowing what was ahead for Mum and ourselves. There is no treatment for CJD, only comfort can be offered to the patient. A rare drug called piracetam had been prescribed to help alleviate the shaking and involuntary jerks but the regional hospital did not have any in stock. When questioned, they told us that they might not be able to get a supply for five or six days. I really had to insist that the drug be supplied that very day and after several assertive conversations with

the ward sister a packet of the drug was found at another hospital about 35 miles away. They asked if I would go and collect it myself! Of course, I got into my car (luckily I have one) and got the drug.

Mum couldn't swallow the tablets that night although the nursing staff begged her to. It was only 24 hours since we had been told of Mum's illness and already it seemed that I knew more about the disease than the staff on the ward. Thank God for the Internet and thank God for the CJD Support Network, whose website was by far and away the most informative. I told the staff that CJD patients have difficulty in swallowing and suggested that the tablet be crushed in lemonade. That is how we administered Piracetam from that point onwards. I stayed with Mum overnight and shared this shift with one of my brothers for the next few days, realising that perhaps this small regional hospital was out of its depth in caring for a patient with CJD. My mother needed an advocate for her needs and I was concerned about that advocacy especially during the night-time hours. The following day Mum was offered a Paracetamol tablet – her prescription had been misread!

By Wednesday Mum had deteriorated so much that she was transferred to a neurological unit some 50 miles away. I had to insist that she was sedated for the ambulance journey as she had developed a fright that is a characteristic CJD symptom. At this specialist unit we were told that a lumbar puncture would be performed to help establish the earlier CJD diagnosis. They said that there are some tropical conditions that can have similar symptoms. It seems strange now but I had this hope that Mum really had malaria – because that can be cured and CJD cannot. A few days later a junior doctor sat me down and said, 'Our tests on the lumbar fluid are negative'. I was delighted and asked, 'If Mum does not have CJD, then

what has she got?’ ‘No, when I say the tests are negative, Mr Bradley, what I mean is that they are negative to anything else’. CJD it is then.

A week since the diagnosis and the family have still not received any advice as to what the future holds or what help might be available for stepfather in caring for Mum. We still have not heard from ‘Edinburgh’ and we are not entirely sure who they are exactly, except that they do some sort of surveillance. In fact, it has taken a week for us to have another brief meeting with the specialist. If it wasn’t for the Internet we would have been very lost indeed.

That first weekend at the neurological unit and I am sitting with Mum when the nurse dispenses paracetamol. Déjà vu does not kick in so I ask what it is for. The nurse says it is Mum’s prescription drug and for a second time I have to point out that it is not.

About a week after arriving here, Mum becomes very agitated indeed and even though she can hardly speak suddenly blurts out a clear request, grabbing stepfather by his collar. ‘Make it go away!’ she says – by which she means her illness. I beg the doctor to sedate my mother but the doctor is most reluctant to do this, saying, ‘We like the patients to find their own equilibrium’. However, in front of Mum we insist on sedation and each night thereafter Mum was sedated to help her (and consequently us) to sleep.

Mum is deteriorating rapidly. Each night when I start the long drive back home with stepfather we ask each other if she can possibly get any worse, knowing that the following day she will have declined yet further.

Two weeks after the diagnosis and we finally receive a visit from ‘Edinburgh’. These two weeks have been a hell for us, so Edinburgh seems to us to be a bit slow. Besides, no help is being offered to us! There is though the matter of a

question and answer session for a survey of theirs so this meeting is more a case of us helping them. At the time I felt angry about this lack of support coupled with a somewhat mercenary need for survey information. This anger was compounded by some basic errors of hospital care that did not seem to accommodate the critical condition of a CJD patient.

I felt angry about this lack of support

The following day Mum’s usual evening medication was not dispensed until midnight. Admittedly, the ward was busy that night but it seems unlikely that there was a patient on it so critically in need of her medication than the shaking jerking frightened woman that my mother had become. We did not leave her until the piracetam and the sedation began to take their effect.

That weekend, Mum continued a decline that was more and more pitiful to witness. I felt that she had lost her eyesight, as it seemed she could not see the utensils when she was being fed. The shaking and the jerking were getting worse, despite the stronger doses of the drug.

Within a week, Mum was transferred to a hospice in her locality. How relieved we were to get her closer to her home and in a quieter, calmer environment. She loved peace and quiet at the best of times. Morphine was administered within a day of arriving and mercifully Mum slipped into a coma. I wondered if Mum would drift back out of the coma but knew that would be cruel, the devastating march of CJD would be continuing and a coma was a much better place for her to be – if not for us. We combed her hair, put her make-up on and even painted her nails. She was the most fastidious of people and would have wanted us to make

sure that she looked her best at all times.

Mum died on Wednesday 23 June 2004 in stepfather’s arms. She just slipped away, her pulse and her breathing just got weaker and weaker. It was the best possible end to the worst possible of all diseases.

The coroner ordered a post-mortem, to be followed by a full hearing. I had been prepared for this possibility in one of many conversations I had been having over the last few weeks with the help-line at the CJD Support Network. I knew that the idea of a post mortem would upset my stepfather so made sure that I was with him when the coroner’s officer arrived. We considered the consent form for the post mortem and made some changes to it.

There is a part of the form that states that photographs can be taken so we scribbled an instruction in the margin that photographs could only be taken of microscopic samples of tissue and not of the body. This is probably what is meant by ‘photographs’ anyway but the form is not specific about its meaning. Mum was camera and publicity shy and she would have insisted on similar safeguards whilst I also know that she would have approved of medical research. Another part of the post mortem form deals with the tricky issue of organ removal, never an easy subject for the bereaved family of a CJD victim. The form states that organs can be removed as part of the investigation but makes no mention of their return. Again, we scribbled in the margin a list of the organs that should be returned to the body after the investigation. That hand written list did not include the brain. We were not giving overt permission for the permanent removal of the brain, but we were not denying it either. This seemed to be the best way of dealing with the issue. Besides which – I rationalized with stepfather – that if the brain were removed from Mum’s body then

the disease would also be removed. Whilst Mum approved of medical research and always used to say that she would like to leave her 'body to science', the decision to allow this is not easy for those who are left to give it.

The coroner's hearing was a long time coming and six months passed. At the hearing the coroner made a number of ill-informed remarks about CJD, which only served to add to our distress at a public hearing. He stated that Mum had died of a variant of 'mad cow's disease'. This was reported on by one of the local newspapers. I wrote to the coroner and pointed out that sporadic CJD was first discovered in the 1920s and that the variant of CJD that has been linked to BSE in cattle was discovered in 1996. Therefore, sporadic CJD is not the variant. Further, I objected to the use of tabloid English in a coroner's court, irrespective of what form of CJD the victim has died of. I asked the coroner not to use this term again if any type of CJD case was being investigated, as it is not medically correct and would not be helpful to any family who are coming to terms with the devastating and cruel death they have been witness to. There was a reply to my letter, apologetic but vague. However, I am delighted to report that we did achieve a small victory at this time. We were able to contact another local newspaper with a much larger circulation figure and a later publication date. Stepfather voiced his concern to the editor about the amount of personal information in the other 'paper's reporting and about our objection to the term 'mad cow's disease'. This newspaper editor was a gentleman of the press, having listened to what stepfather had to say he agreed to print a version of the story that was acceptable to us and which would've been more acceptable to Mum.

So why write a brief history of my mother's dismal experience? I write only with a spirit of constructive criticism, in the hope that the care and support of a person

with sporadic CJD can be urgently reviewed and improved. Also, I can make some recommendations that would improve the support offered to the victim's carers. First, in this modern age, a family told of this terrible diagnosis should be assigned a support worker immediately – there and then. This person could be based remotely (the rare nature of CJD might dictate that this would be the case) and would be required to contact the family using the telephone (it's what mobiles were invented for) on the day that the diagnosis is made (even if that day is a Sunday and the time is 8 pm.)

A family told of this terrible diagnosis should be assigned a support worker immediately

The specialist making the diagnosis should be required to gather the contact details of the next of kin and these should be passed to the support worker's team on a 24-hour national number. In other words, carer's support should be 'on call'. I would like to see help, advice and support becoming immediately available. Not everybody has access to the Internet and even those that do may not find the information rich repository that is the CJD Support Network website. A support worker could make sure that information and options for care are immediately available. More importantly, such a worker could answer the numerous questions that will obviously arise on an ongoing basis.

Secondly, a similar level of support and instant advice should be made to the nursing staff that will be caring for the patient. This action should also be the responsibility of the support worker's team.

In all cases, I cannot over emphasize the need for urgency. The alarming deterioration of a person with CJD cannot wait for drugs to be found and delivered and a family should not have to wait for weeks

for their questions to be answered. In 2005, in a developed country like ours there really can be no excuse.

Personally, I decided to get involved with the CJD Support Network – though this was difficult so soon after my mother's death. During my mother's illness we learnt a few things that might help to improve the comfort of the patient and no doubt other carers learnt good practice also by default. As a totally non-medical lay person, I have decided to compile practical advice in an easy to read booklet so that others who are plunged into this awful situation will have a better starting point. For instance, we learnt very quickly that Mum could not decipher the patterns on the clothing we were wearing and could in fact become frightened of them. So we decided to wear plain clothing at all times. Also, we found that switching her bed side light off at night did not help her as she became alarmed in the dark (I learnt that this is called 'sun downing') and so we always left her light on just in case she woke up and needed to see where she was. It's little things like this that can make a big difference to the comfort of the patient.

If you have experiences like this that you would like to share please contact me via the CJD Support Network (or email me at: bradley449@yahoo.com).

If you need any help or information about any type of CJD please contact Gillian Turner at the CJD Support Network help-line. Gillian provided me with a support that I passed on to my family and hence on to my mother during the terminal stages of her illness. This made an immeasurable difference to my mother's standard of care. The marvelous work that is undertaken by the Network always requires more money. If you have any money-raising ideas please contact Gillian. To all people who have been affected by this terrible illness I offer you my wholehearted support and solidarity.

Fighting together as a family

By Mike

My wife Joyce always joked that she hated reaching fifty and how true this was as first cancer and then CJD attacked her body.

The summer of 1999 began with the loss of Joyce's best friend from breast cancer, the shock, or so we thought, found Joyce suffering with severe backache. Treatment from an osteopath proved fruitless and with the problem worsening, Joyce was referred to the hospital for tests. Numbness had spread to her legs and upper body and tests revealed a tumor on her upper spinal column. A biopsy showed that the tumour (lymphoma) had been malignant and a course of radiation followed to kill any remaining cancer cells. Physiotherapy was necessary following the paralysis to help her walk again and hospital visits became a way of life. Ten months later she could drive her car again and life seemed to be back on track, although now she had constant back pain.

But the good times did not last. Ten months later in May 2001 when we were on a family holiday in Spain she complained of some deafness in both ears. Initially we thought it was just wax caused from the water whilst swimming, so a visit to the practice nurse was arranged. This was not the problem so an appointment was made with the doctor who again referred us back to the cancer hospital in view of her previous problems. Various tests were undertaken over the next three months without any solutions and by September she could hardly hear anything and was beginning to have trouble reading and remembering words. She was then referred to another hospital and admitted for further tests in a neurology ward. During both her

illnesses Joyce had remained strong but now – with the world being cut off to her, unable to hear, limited reading and writing skills, one could sense how frightened she was not knowing what was happening to her.

We were always a very close family and together we helped her through these changes and supported her with love and affection

After nearly three weeks of tests, including a brain biopsy operation, there was still no diagnosis, so the consultant suggested we go home and come back in a month. During this time the condition worsened slightly and when we again met the consultant a lumbar puncture and blood tests were undertaken and he indicated another brain biopsy may be needed if these tests did not reveal anything. That night everything changed. When Joyce woke up next morning the caring and loving woman I married had disappeared and what remained was a confused and anxious woman.

She no longer wanted to leave the house without being accompanied by myself and the children. At home she still wanted to cook and clean for the family but could no longer remember how to use things such as the washing machine. Now she only knew the recipe for one meal which she wanted to cook for us each day. She was continually confused and had short memory recall which naturally caused her to get very upset when we could not understand what she wanted.

With life confused for her she could not understand things such as why our daughter did not go to school at weekends. Her memories were of past events and even her taste in food and drink changed back to when she was in her early twenties. The children and I were devastated. Why had this happened? Until now we had hoped that the doctors would find a cure for her; but the illness had changed and things were to get worse over the coming month. She could no longer write and began speaking less and less, now needing support to dress and encouragement to eat meals. We were always a very close family and together we helped her through these changes and supported her with love and affection which seemed to make the situation bearable for us all. We were aware of what had changed for her and could only imagine how scared she was inside.

It was late December now and as the previous tests had proved fruitless the hospital indicated they would like to do a second brain biopsy operation. I could see little point in putting her through the operation again but you still had hope for some miracle cure and I agreed for it to be done. However, by the time of the operation she did not want to leave our home. The first two attempts to go to hospital were unsuccessful, but with the help of my son and daughter I managed to get her there, but she was scared and the doctors had to medicate her on arrival. I spent two days and nights at her side and it was only after we left the hospital that she again relaxed. This time the consultant had arranged for an MRI scan to be undertaken before the operation so that the brain biopsy

could be more focused. Three weeks later, after eight months of tests and hospital visits, the consultant had the answer.

In a room full of his medical team numbering about twelve people the consultant told me my wife had the terminal condition of Creutzfeldt-Jakob disease and handed me an information sheet containing details of the illness and contact numbers. He told me that the Prion Unit at St Mary's Hospital would be in contact shortly and that for my own peace and mind it would be better not to search the Internet for information and wait and see them.

I waited a few days but needed to speak with someone and contacted the CJD Support Network. We spoke for well over an hour and how reassuring it was to be able to talk about the illness, its effects and what support I should be able to receive. The next day their handbooks and literature arrived in the post and I was able to read about everything about the illness. As with anyone in my position I wanted answers; how did she get it, why, what will happen and most importantly how long does she have. In truth these answers do not exist, but you do not know that at the time and cannot accept the uncertainty. It is only time and a lot of conversations with friends, family and the professionals that help you to cope and provide the necessary support to your spouse or family member. That is how I managed to cope with this difficult situation, but in many ways I have been lucky as my wife is still with me after four years of suffering with this illness, although I know in my heart she would never have liked to survive in this way.

A week later I contacted the Prion Unit and arranged a meeting with Professor Collinge. He went through the illness in detail with me and the children and explained the various forms and how things might progress with sporadic CJD

which had been found in my wife's brain biopsy. Afterwards, one of his team recapped the main points and answered any further questions we had. They provided some more contact numbers, including one for the Admiral nursing service in my area who would support me as the main carer.

Only time and a lot of conversations with friends, family and the professionals help you to cope

The hospital passed our case to the care manager from my local medical services. She visited me and my wife at home and went through everything to do with the support available to us. She was keen to find out what help we needed and to get things in place quickly. The first thing was to get her colleague, the operational therapist to make contact. He called to see us a day or two later, clearly he had done his homework on the illness and knew what would be needed for me to keep my wife at home. He covered everything thoroughly with me and I made him aware of the special social services guidelines concerning this illness which had been provided to me by the CJD Support Network. From the guidelines he realised that the care coordinator at the Edinburgh centre needed to be involved should there be any funding difficulty by my local authority and made contact on my behalf.

The care coordinator from there contacted me direct and arranged to come and see us. We had an extremely useful meeting a few days later when we went through everything again to do with the illness, what might happen and how social services will support us. A further care meeting was fixed so that various units within the social services for the area could attend and these included our doctor, the

care manager, operational therapists, district nurses, palliative care team, community physiotherapists, speech and language team, wheelchair service and the Admiral nurse. At that meeting the care coordinator explained what support would be needed and gave guidance on how each section of the social services could help.

Over the next couple of weeks all those who attended made contact with me and the support started to be put in place. The care manager provided temporary assistance from their home help team with washing and dressing my wife and arranged for me organise myself a carer under the council's independent living scheme. This would enable me to have some help when I needed it. This suited my needs as I preferred to wash, dress and feed my wife. The operational therapists provided all the equipment – commode, bathing aids, hoist, slings and numerous other things. We discussed how a downstairs bathroom would help with washing when my wife could no longer walk and he provided guidance on how to tackle this and seek the necessary financial support. Four months later my wife would be unable to walk and living downstairs. Fortunately the bathroom was in place by then and this has enabled me to shower her each day which you can see she enjoys. The operational therapists as well as the care manager visited regularly over the early months of the illness and provided me with both practical and moral support I needed.

Indeed I would like to say that everyone I have been in contact with in social services during my wife's illness has been extremely helpful and supportive. The district nurses and palliative care nurse have kept regular contact throughout the illness and I know they can be contacted, together with our GP, whenever guidance or help is needed. When swallowing problems occurred, someone from the speech

and language unit came out at short notice and provided the advice and reassurance I needed. The community physiotherapist visits at regular intervals to check on my wife and has given me guidance on the exercise movements needed and help with tackling her neck support. When the wheel chair broke and could not be repaired a replacement was purchased and delivered within a few days. The Admiral nurse visited regularly for the first year of the illness and her support with listening and talking was tremendous therapy for me coping in those early months.

I have also been lucky in that my employers gave me the time I needed early on to be with my wife and family and the financial security to continue caring for her. This together with the social security benefits package and care support allocation has enable us as a family to adapt to the predicament and provide my wife with the best possible care for her remaining time.

My wife's illness has not progressed as with many other cases of sporadic CJD. Her hearing returned after six months, which has enriched her world. Early on in the illness when she lost the ability to speak she would respond to us with facial expressions and noises but now this is not possible and she reacts using her eyes and, on a very good day, a noise as well. She has not been able to walk for over three years and cannot purposely move her arms and hands. Initially she ate finger food, but it was not long before I had to feed her and the food needed to be liquidized. There has naturally been some weight loss and muscle wastage but with a regular routine and healthy diet she has kept reasonably well with the main changes seen in the different levels of dementia and swallowing,

Joyce never liked taking medication and throughout the illness I have tried to keep it to a minimum. We use a suction

machine to support her swallowing and the usage depends on how she is that day. Additionally we use atropine drops morning and night which thickens the saliva and this normally helps to reduce swallowing problems. I have found rubbing 'Vicks Vaporub' on her throat and the edge of her nostrils helps to generally relax Joyce when she is tense and has swallowing difficulties. Olbas oil is also beneficial for this problem and for 'spasms' that attack the body. The only other medication is a low dose of half a teaspoon of clonazepam at night which relaxes the body and helps her sleep. I believe that talking to her at all times gives the reassurance she needs and acts as the main medicine.

Everyone I have been in contact with in social services during my wife's illness has been extremely helpful and supportive.

A normal day starts with a shower and some brief exercises which provide movement and stimulation for her. She is then dressed and given her breakfast. This is normally a banana and kiwi which has been liquidized with multivitamins syrup added, followed by one or two yoghurts and then 200mls of cranberry juice mixed with summer fruits squash and thickened to same consistency as the other foods. Usually weather permitting I take her out for a walk in the wheelchair for 30/40 minutes which again acts as stimulation for her. If this is not possible she sits in her wheelchair anyway and this gives her a change of view other than the bed.

Lunch always consists of potatoes, carrots, and cabbage or brussel sprouts with chicken, mince or fish and again the meal is liquidized. She has a pudding of trifle and custard or one of the

many flavours available of instant whip followed with a thickened barley drink. Every day she listens to her favourite music using a CD and headphones. Her bed is situated in our living room so the everyday comings and going help to keep her stimulated. The evening meal is very light and consists of a barley drink and yoghurt or instant whip. I use a little water to wash her mouth after every meal and remove any excess with the suction machine.

Initially with the illness we would have four very good days each week when my wife was bright and responsive. Then there would be a couple of bad days with spasms, swallowing problems, deep dementia, while others in between became an average day. Gradually the good days disappeared and every day has become average with good and bad moments. You learn to take one day at a time and realise you need support if you are going to cope. No longer do I have the same energy levels as I did when I started caring for my wife. I rely on the help of the two carers who work a few hours each weekday; the two ladies who sit with my wife now for five nights each week, and my daughter and son, who often get involve when I need help.

My wife's condition continues to slowly deteriorate but I know she is comfortable in her home surroundings, reassured by the familiar voices around her. I am sure it is this together with the love of her family and a few close friends which has given her the strength to fight this illness. Eventually the CJD will win, but until then we will fight together as a family.

EDITOR'S NOTE: Since Mike submitted this article, sadly, Joyce died peacefully in his arms on the 29 November 2005. All his friends at the CJD Support Network send their sincere condolences to Mike and his family.

Alan's story

Told by Florence, his wife and carer

Alan and I met in 1966 and married in 1967. I was 19 and he was 23. He was a keen cricketer, a semi-professional footballer, a keep fit enthusiast, a marathon runner and a five-handicap golfer. Having left school at 15 with no formal qualifications, he was nevertheless ambitious, with a tenacious spirit and determined to better himself in life. By 1973 we had two lovely sons, Edward and Richard.

Alan was an extremely hard-working, fun-loving character who lived for sport and his family and had built up an international sportswear company. By 1997 we moved to Cornwall where we operated a very busy city centre restaurant.

In October 1998 he became increasingly forgetful and by December 1998 he was extremely anxious. I attributed this to the stresses of a new business venture and felt sure that a January holiday would recharge his batteries. We went to the Algarve, a favourite destination for us of some 20 years, to play golf and to invest in a retirement property. He became more anxious and impatient and would complain that we had not been active enough; so I would get myself ready for a walk only to find him fast asleep on the bed. In a restaurant he was unable to decide his order and became irritated with the waiter.

On our return in early February 1999 he complained of a severe headache, and by then was taking daily headache relief medication. Alan was not a complainer. He had seen the GP once or twice during the year complaining of poor vision, and an eye test and spectacles were recommended, which he hardly used and said he did not need.

A visit to the doctor was a rare occurrence for Alan, and at this

point I became increasingly worried. He had become withdrawn, was unable to function at work and would sit for hours.

Alan was an extremely hard-working, fun-loving character

A visit to the GP resulted in a prescription for Prozac, an anti-depressant to counteract his low mood. By the following weekend he was more withdrawn and his gait had become unsteady. During the next two weeks Alan was seen by a psychiatric nurse. On 1 March he was seen by a registrar at the local psychiatric unit who admitted him to the psychiatric ward after an hour of questioning. I visited him daily and was horrified to witness the deterioration in his condition. By the second evening he was unable to feed himself. My concerns were met with disbelief and I had to push hard that week in order to get the registrar to action any investigation.

By Friday a brain scan was organized and the result showed 'no tumor'. *I was assured that this was good news* and yet my husband could barely talk, and was incontinent, having been left on a chair in the corridor of the psychiatric unit. My alarm bells rang, and after many phone calls, and at my insistence, he was transferred to a neurological ward in a nearby hospital on 5 March. There was no consultant available to assess him since it was Friday evening.

During the next two weeks he underwent exhaustive tests for numerous diseases from Aids to Wilsons disease, including an EEG and a lumbar puncture,

which was excruciatingly painful. By this time he could no longer speak.

On 15 March the consultant informed me that Alan had gross abnormalities in the brain, but no clear diagnosis was given. I was devastated but determined to nurse him; no matter what; and planned in my sleepless hours all manner of practicalities to this effect.

On 17 March I was given the diagnosis of classical CJD and informed that the team from the surveillance unit in Edinburgh would come to assess Alan and confirm the diagnosis. The consultant told me to expect the worst and that Alan had only weeks to live and that I would need to consider decisions about continuing to feed since he would very soon lose the ability to swallow. By this time Alan was unable to see, but had a fixed look of terror in his eyes. I stayed most of the day and during the night in his room to help to calm him during his fearful hallucinatory bouts of aggression which were traumatic to witness. I made plans to nurse him at home, but soon realized that the complex, and intense nature of care required was impractical and impossible.

By 23 March, Alan was admitted to a hospice some 20 miles away where I was able to stay some nights. I was determined that, if nothing else, Alan would have some peace and dignity, but his deterioration, even with anti-convulsive drugs, was the most traumatic experience I have witnessed.

Alan died in my arms on 11 April aged 55, having fought hard not to leave his family.

Funds have been raised in memory of Alan, to fund a new leaflet for the CJD Support Network. See page 2.

Blood donors who gave to vCJD cases

a new group of individuals notified as 'at-risk' of vCJD

Dr Kate Soldan, Health Protection Agency

Why might blood transfusion be a risk for vCJD? The risk of vCJD being transmitted by blood transfusion has been a concern in the UK for a number of years. The main reasons for this concern are:

- evidence from animal studies that vCJD can be transmitted between animals by blood
- evidence from studies of human tissues that infectivity for vCJD can be found fairly widely throughout the human body
- vCJD infection has been observed in two recipients of blood transfusions from donors who later developed vCJD.

Precautions

What has been done about this risk? A number of precautions that have been taken as a consequence of this concern, including:

- A number of steps taken by the Blood Transfusion Service to reduce the risk of vCJD infection in blood transfusions (eg selecting donors with no risk factors for CJD, removing white blood cells from blood)
- Stopping the use of plasma donated in the UK in the manufacture of medicines from plasma
- Tracing certain people who can be identified as 'at-risk' of vCJD because of blood transfusion (eg from a donor who later developed vCJD) and asking these people to take some precautions in order to reduce the risk of vCJD being passed from patient to patient during medical care.

In the summer of 2005, this last precautionary measure was extended to include some people

who have donated blood to patients who later (some time after their transfusion) developed vCJD.

Why? How many cases of vCJD are linked to transfusions? Over two million blood donations are collected each year by the UK blood services and in excess of half a million patients receive transfusions annually. Of the 151 people who have died from vCJD in the UK (to 4 October 2005), only four have been confirmed as having received blood transfusions that may be associated with their subsequent development of vCJD. (For two other cases, symptoms developed before or very shortly after transfusion and, therefore, transfusions are not considered a possible source of their infections.) For one of these cases, the probable source of infection has already been identified as one of the donors went on to develop vCJD. For the remaining three cases, it is not possible to know whether they were infected with vCJD by their diet (as is suspected for most vCJD cases which have not been transfused) or by blood transfusion. It is also not possible to know which – if any – of the 110 blood donors who gave blood to these cases might be carrying vCJD infection.

Risk assessment

What has been done about these donors? A risk assessment was conducted to estimate the probability of these donors being the source of vCJD infection for the three cases. An expert committee, the CJD Incidents Panel, then considered this risk assessment and recommended what should be done.

As a precautionary measure, all these donors have been traced by the blood services, and are being

advised by their doctors about the precautions they should take. These individuals are being informed that they are 'at risk of vCJD for public health purposes' so that special public health precautions can be taken to reduce the risk of person-to-person transmission of vCJD during their healthcare. These precautions are:

- not to donate blood, organs or other tissues, and
- to inform their healthcare providers of their 'at-risk' status so that some special infection control guidance can be followed for the instruments used on them for certain medical procedures.

The blood services, the Health Protection Agency and the doctors of these patients are working together, and with other experts, to try to answer the questions that remain about these donors, and about the risk of vCJD associated with blood transfusion.

What happens next?

This notification of 'donors to vCJD cases' was one further precautionary measure to reduce the possible risk of further transmission of vCJD in the UK. Only time and further work will tell how well this has worked. The individuals involved will be told if anything is learnt that changes their situation. Work (eg other risk assessments, and expert meetings) is continuing to try to identify whether there are other groups of patients for whom precautions would also be worthwhile.

Further information about this and other notifications of patients, can be found at http://www.hpa.org.uk/infections/topics_az/cjd/menu.htm.

Prion protein accumulation in UK tissue samples

A survey of appendix and tonsil tissues from anonymised individuals found three positives out of 12,674 tested. This gives an estimated prevalence of vCJD infection of 273 per million in the UK. This figure is higher than the current cases of vCJD would suggest. It might, however, indicate that there could be more asymptomatic infections, with a 'carrier state' that could result in secondary transmission via blood or surgical instruments.

More study is underway to see if there are any genetic differences which might explain the above findings.

For further information on this study see: 'Prevalence of lymphoreticular prion protein accumulation in UK tissue samples' Hilton et al; *Journal of Pathology* 2004; 203; 733-739.

CJD Support Network Management Committee 2005-06



Dr Angus Kennedy
– Chairman
Consultant Neurologist



Maria Bryne – Secretary
Maria is a mother with three children whose husband Graham died with GSS



Mike Curtis – Treasurer
Mike, a former bank employee whose wife, Joyce, recently died of sporadic CJD



John Gilbert – Assistant Treasurer
John's brother in law died of sporadic CJD



John Williams
John is a retired Government engineer whose daughter, Alison, died of vCJD



Stuart Durkin
Stuart's father died of sporadic CJD in August 2003. Stuart is a freelance social researcher



Anita Tipping
Anita is a state registered nurse, RSCN, whose son David died of CJD through growth hormone injections



Michael Bradley
Michael's mother died of sporadic CJD



Jean Lightband
Jean is a neurological specialist nurse at the National Hospital for Neurology and Neurosurgery



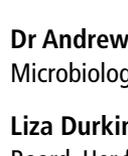
Roger Tomkins
Roger's daughter Clare, died of vCJD



Sarah Tomkins
Sarah's husband Edward died of sporadic CJD



Derrick Biggs Derrick is our social services adviser and an operations manager with Cambridgeshire Social Services. Derrick is the Association of Directors of Social Services link person for CJD



Dr Andrew Smith Andrew is a Senior Lecturer in Microbiology at Glasgow Dental School



Liza Durkin Liza works for the Youth Justice Board. Her father died of Sporadic CJD



Gillian Turner
CJD Support Network co-ordinator

IMPORTANT DATE FOR YOUR DIARY

International conference on CJD Decade past – decade to come

Monday 20 March 2006 · BMA House, London

The past decade has seen new challenges, knowledge and hope for a future treatment for this devastating disease. Will these be realised in the next decade? It is our plan to bring together professionals, families affected by all strains of CJD and those new people who have been informed that they are at a higher risk of CJD through secondary transmission.

To register your interest, contact CJD Support Network, PO Box 346, Market Drayton, Shropshire TF9 4WN. Telephone 01630673993. Email info@cjdsupport.net. Or visit www.cjdsupport.net.

The CJD Support Network is now recognised as the leading charity supporting families and professionals affected by all strains of Creutzfeldt-Jakob disease.

Talks include:

- **Fundamental understanding of what prion disease is** – Professor John Collinge, Director of the National Prion Unit,
- **Epidemiology and risks** – Dr Richard Knight, Director of the National CJD Surveillance Unit
- **A decade of pathological discovery in CJD** – Professor James Ironside, Consultant Neuropathologist at the National CJD Surveillance Unit
- **An overseas view of treatment issues** – Dr Paul Brown (USA)
- **An overview of DOH CJD committees** – Professor Don Jefferies
- **The work of the CJD Incidents Panel** – Dr Nicky Connor, Health Protection Agency
- **Blood risk past and present** – Dr Pat Hewitt, National Blood Service
- **The public and patient perspective** – Harry Cayton, Director of Patient and Public Involvement at the Department of Health