CJD SUPPORT NETWORK

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Newsletter

Issue 7

March 2000

CREUTZFELDT-JAKOB DISEASE SUPPORT NETWORK NEWSLETTER

NvCJD renamed vCJD

At its meeting on 18 March 1999 the Spongiform Encephalopathy Advisory Committee (SEAC) agreed that 'variant CJD' (vCJD) should be used in preference to 'new variant' (nvCJD). This is in line with current practice in many scientific journals. 'vCJD' or 'variant CJD' has been used throughout this newsletter.

BSE enquiry extended

The BSE Inquiry has been extended for a further six months. Lord Phillips said: 'We want to produce a clear and readable account of the BSE story that sets out our key findings, conclusions and recommendations ... Many have devoted enormous time and energy to providing us with evidence and submissions. If our report is to do justice to them, it cannot be produced in a rush.' The report will now be submitted to the Government by 29 September 2000.

On other pages

CJD memorial service

A memorial service for people who have died from CJD was held by the Network at St Martin-in-the-Fields, Trafalgar Square on 12 December 1999. Specially chosen hymns, prayers and readings reflected the many concerns raised by CJD. A congregation of 150 people, including the families of many of those who have died, attended the service which was conducted by the Rev Nicholas Holtam. Readings were given by Patricia Nolan, whose mother had died from classical CJD, and Michael Sinnott, whose daughter had died from vCJD. John Williams and Margaret Conan read out 78 names of people who had died and candles were lit in their memory. The service concluded with a cello recital by Michael Evans, made possible by a charitable grant from the Academy of St Martin-in-the-Fields.

After the service a reception with mulled wine and mince pies was held in the crypt. A collection of almost £500 was divided equally between the church and the CJD Support Network. The donation to the Network will be used to fund the caring grant scheme which helps families in need. Copies of the service sheet and memorial role can be obtained by writing to Gillian Turner. (Memorial address: page 2)

Summary of address at the CJD Memorial Service

Prof Sir John Pattison, Director of Research and Development at the Department of Health

I feel honoured to have been asked to address this memorial service. I also feel somewhat inadequate to the task. I do not have your own direct involvement with family members who have died of CJD. I have not had first-hand experience of the dismay at the onset of signs of the disease, of the anger at the apparent failure of many people to recognise what was happening and act accordingly, and of the despair when trying to answer the question 'why us?' and reflecting 'what a waste'.

The deaths of your loved-ones have all been inappropriate and in some cases very untimely. Nevertheless, what has happened to them has focused the attention of many and has led to action. The existence of the CJD Support Network and the fact that this memorial service is taking place are expressions of that. More widely, whole governments have acted and continue to act. During the coming year there will be an official report on one form of CJD and its origins, after a very extensive and public inquiry. I do not know what it will contain but have no doubt that it will try to understand what happened and develop ways of ensuring, as far as possible, that such a thing will not happen again. This inquiry came about because of what happened to your wives, husbands, sons, daughters and friends.

It is an unfortunate fact of life that it is after tragedies that the most intense demands for answers and explanations come. So it has been with CJD and similar diseases. CJD was first described about 70 years ago but it is the events of the last ten years that have resulted in intense interest in a disease formerly referred to only in highly specialised medical texts.

The search for answers is progressing, though much more slowly than any of us would wish. The descriptive phase of the disease, which is always the first phase in developing an understanding of what is going on, is all but over for CJD. There has been some progress in understanding the mechanisms by which some of the abnormalities associated with the disease are produced. This will help us make progress in finding diagnostic methods and ways of halting or even reversing the disease progression. It would be wrong to give the impression that we are near a breakthrough of any sort, or that it will not take many more years to achieve this: CJD is a disease which has some features that are quite different from all others.

One other question that you may be asking yourself is: 'Why us?' Why was it that our relative was affected by CJD and yet the majority of others were not? This is a question that is difficult to answer. There are great similarities between all men and women, yet the relatively small differences between us make us instantly identifiable as individuals. Some small individual differences in genetic makeup and lifestyle will make us more or less susceptible to disease. It is these small differences that combine to determine who contracts a particular disease. In the case of CJD we may never know the precise details involved.

This memorial service has power and significance by virtue of people coming together to share a loss. However, it is your individual memories of your relatives and friends that will be most powerful. I cannot share that with you directly but I can join you in recognising that through their lives, and through their deaths, they have changed the world for us all.

Evaluation of the CJD Support Network

A summary of the findings and recommendations

The Network's application for three-year funding to the Department of Health in June 1996 included a sum for an independent evaluation. The evaluation had three aims:

1 To provide an independent evaluation of the formation of the Network against the objectives set for the project.

2 Consider how the evaluation might point towards the future of the Network.

3 Assess if the Network could provide a model for the establishment of similar support support for other dementias.

The evaluation, undertaken by Des Palmer, concluded that the Network is in good shape. It has secure funding to meet its current level of expenditure until April 2002. It is protected and nurtured by a larger, well-resourced and highly respected national charity. It has a reasonably sized membership and mailing list, a clear sense of purpose and a committed management committee. It provides a range of services to families and to relevant professionals. It is actively linked into a network and, in some respects, is the linchpin. In the relatively small field in which it operates it is valued and respected. That it has come so far in just five years is a major achievement.

The report makes 21 recommendations to improve and develop the effectiveness of the Network. These recommendations will be considered carefully and implemented within the resources available.

Copies of the full evaluation report can be obtained from Clive Evers, Chairman, CJD Support Network, Gordon House, 10 Greencoat Place, London SW1P 1PH. Please send a large self-addressed envelope with a 39p stamp.

Annual General Meeting

The fourth annual general meeting of the CJD Support Network was held at the national office of the Alzheimer's Society on 2 October 1999.

The current committee was re-elected:
Chairman: Clive Evers
Secretary: Margaret Conan
Treasurer: John Gilbert
Members: Maria Byrne, Patricia Nolan, Anita Tipping, John Williams

Professor Bob Will, of the National CJD Surveillance Unit, and Kathryn Prout, St Mary's Prion Clinic, were co-opted onto the committee.

Minutes of the meeting are available from Gillian Turner, address on the back page. The next AGM wll be held on 7 October 2000 at a venue to be arranged.

In memoriam donations

The CJD Support Network gratefully acknowledges the following in memoriam donations recveived over the last six months. All such donations are put towards the Network's Caring Grant Fund which is used to provide direct help to carers of people with all forms of CJD. This help is much appreciated.

In memory of: Michael Morgan; June Chadderton; James Arthur Cox; Pam Watson; A Broadhurst; Barry West; G Day; Marianne Harvey; Neville Price; R Steel.

INFORMATION

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Cycling round Blackpool

Jason Richardson will cycle the Tour of Blackpool this spring in aid of the CJD Support Network. The son of Jason's work colleague, Irene Smith, died with vCJD in November 1998. All funds raised will be used to provide caring grants to families who are looking after someone with CJD.

To sponsor Jason, contact Gillian Turner, national CJD case co-ordinator, on 01630 673993, or write to Jason at 32 Kershaw Crescent, Luddenden Foot, Halifax HX2 6NS.

London Marathon

Matthew Southwell of Hebden Bridge will be running the London Marathon in aid of the Network. Contact Matthew at 1 Stubbing Brink, Hebden Bridge, West Yorkshire HX7 6LR.

Specialist testing for the rogue protein involved in vCJD

Tonsil biopsy

Professor John Collinge

Difficulty of diagnosis

Diagnosis of vCJD can be difficult, particularly in its early stages. While there is no treatment at present, it remains very important to reach a clear diagnosis as there are a number of other treatable diseases which cause very similar symptoms. 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 much less clear. In certain situations, therefore, a brain biopsy may be considered.

Infected defence system

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 so-called '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. We have shown that by removing a small piece of tonsil tissue in appropriate patients it is possible to detect the rogue form of prion protein thought to be the infectious agent (or prion) that causes CJD. The tonsil test is only positive in vCJD. It does not diagnose other forms of CJD.

What does the test involve?

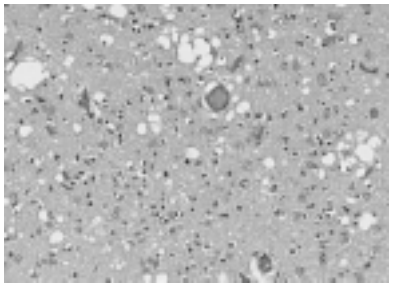
At present, tonsil biopsy is only performed at the Prion Unit at St Mary's Hospital, London which has specialised facilities for patients affected by prion diseases. Patients are normally referred to this service by consultant neurologists around the UK and they will usually have already undergone extensive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

The patient is assessed over a three- to five-day period. This assessment will recommend whether or not a tonsil biopsy should be performed. Some patients will have had their tonsils removed (tonsillectomy) during childhood but usually sufficient tonsil remains to do a successful biopsy. The test requires a general anaesthetic, so the patient will be 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 and is a very simple procedure. A special disposable biopsy kit is used. The patient normally stays in hospital overnight to ensure all is well. St Mary's is a busy teaching hospital and, like all NHS hospitals, there are often pressures on availability of beds. The patient may therefore sometimes need to wait a little longer for a biopsy.

Although devastating, a firm diagnosis of vCJD ends uncertainty

Once the biopsy has been taken, two separate types of test are performed. One involves looking at the tissue with a microscope after treating it with a special stain to detect the rogue form of the prion protein. The second test involves detection and characterisation of the protein in a special laboratory, 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

Microphotograph of spongiform change in brain tissue taken from someone with CJD



Background to CJD

There are a number of forms of CJD but only one of these, first described in 1996 and now known as variant CJD or vCJD, is linked to exposure to BSE. Unlike classical CJD, vCJD predominantly affects young adults and so far remains a very rare condition. 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.

biopsy and this is either communicated to the patient and their family by their local doctor or at an outpatient clinic.

Need to know

Although devastating, a firm diagnosis of vCJD ends uncertainty and removes the need for doing any further tests which might otherwise be considered. Establishing a diagnosis is also extremely important in developing an individualised package of care 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 and develop such a test.

Further information

Informal enquiries to can be made to Kathryn Prout, Prion Nurse Specialist. Tel: 020 7886 6883 (answerphone). Email: k.prout@ic.ac.uk. Website: http://www.med.ic.ac.uk/dn/dngm/prion-clinic/clinic.htm

Alternatively a neurologist, GP, geneticist, psychiatrist or any other doctor wishing to refer a patient suspected of having any form of prion disease should write directly to: Professor John Collinge, Consultant Neurologist, Prion Unit, Department of Neurology, St Mary's Hospital, Praed Street, London W2 1NY.

CJD in the news

Guidance on CJD updated

The Department of Health has issued two circulars reinforcing advice on CJD. 'Washing and decontamination of medical devices is the single most important step in reducing the risk of transmission of infectious agents, and is a key measure in minimising any risk of vCJD,' said deputy CMO Dr Jeremy Metters.

The text of these circulars is available on the DOH website: www.open.gov.uk/doh/coinh.htm

Hidden scars

The New Scientist (21 Aug 1999) reported that there may be an easier way to spot vCJD. Alan Coulthard and colleagues at the Royal Infirmary of Newcastle upon Tyne say that it may be possible to diagnose vCJD using an MRI scan. Deep scarring in the posterior thalamus in the brain only appears to occur in vCJD. Martin Zeidler (formerly of the CJD Surveillance Unit) says that current research is 'pretty impressive' but that further studies are being carried out to determine the reliability of the tests.

Predicted numbers

There have been a number of published articles regarding the predicted number of cases of vCJD. Many simply state that the number of victims is still unknown. The Daily Mail, dated 22 September 1999, quoted the Government's chief medical officer who stated that the possible range of cases was between a few hundred up to several million. On the last day of the BSE Inquiry, Judge Phillips said that the numbers seen so far could be 'just the tip of an iceberg'.

Researchers at the Wellcome Trust in Oxford suggest that the number of cases in 1999 and 2000 may be a good indicator of future cases. If the final figures for 1999 are 15 cases or fewer, their prediction is for a maximum of half a million cases in total. If the year 2000 shows a decrease, or at least no increase in cases, Wellcome predicts 14,000 cases or considerably fewer. Scientists at a number of centres are also screening thousands of tonsil and appendix tissue samples to determine the presence of vCJD. This will enable them to gauge the prevalence of the disease in the population as a whole and thus to be able to make a better prediction of total future victims.

Protein that could block CJD

The Lancet (Vol 355, 15 Jan 2000) reported that a team of scientists, led by Claudio Soto, are closer to finding a possible treatment for vCJD and similar diseases. They have found a peptide (a protein component) which can stop and reverse the damage caused in the brain by CJD. So far, this has only been carried out on laboratory mice.

vCJD = BSE

Powerful new research by Professor Will and colleagues, reported in all the daily newspapers in December, proves that vCJD and BSE are 'indistinguishable'. Proc Nat Acad Sci (PNAS), 1999, 96, pp 15137-15142.

CJD study day for social workers and social services staff

The aim of this study day on 1 November 1999 was to inform social services professionals about the clinical features of CJD, the role of the social worker, the process of assessment and innovative ways of funding care packages.

The role of social services

Derrick Biggs, operations manager for Bedfordshire Social Services, said that social services must act very quickly in cases of CJD. A keyworker should be appointed for each new case and services should be co-ordinated and information shared amongst professionals and families.

People should be informed of the likely response times and there should be a full explanation of the process of an assessment and what comprises a care plan. The care plan needed frequent updating – not just once every six months.

Delivering services should be undertaken with sensitivity. Trust needs to be established with carers. Imagination and continuity were two vital elements of the services agreed in any care plan. The possibility that a patient may die at home should be acknowledged and appropriate links and support from community nurses was vital.

Points raised in discussion included the possible need to involve a welfare rights worker early on and the importance of responding to concerns from relatives about infection control, particularly where there were children in the household.

CJD guidelines

Kate Register, planning and policy development officer from Bedfordshire Social Services, explained how the department had adopted the ADSS/CJDSN guidelines on CJD.

Physical disability teams were nominated to take up CJD referrals and, in December 1998, an outline policy statement was drafted and subsequently distributed to team leaders and named individuals in the disability resource centres and information services.

Two half-day workshops were run by the CJD Support Network in May which were attended by 18 staff from all the different teams in the county. There were representatives from the community mental health team, the over-65 team, the learning disabilities team and the Pathfinders team which specialised in the care of people with dementia.They found the training worthwhile and had subsequently had to respond to two cases of CJD.

Assessment and case management

Alex Hartshorne explained the process and components of care assessment and case management. There were seven key stages of care management: gathering information; deciding on the level of assessment and appropriate referrals; assessing the need; reviewing the assessment; planning the provision; implementing the assessment; and monitoring. She explained some of the key legislation, which empowers social services to carry out assessments, including the 1990 Community Care Act section 47 and the Carers Act 1995 which stresses an individual's right to an assessment.

She reminded delegates that, while the process of assessment is regarded as important by professionals, carers are most concerned with its outcome.

The need to involve the patient's GP was crucial, as was the requirement for more training and education on neurological illness for those working in primary care. It was also important to enable good practice to be identified and passed on to other professionals.

Clinical presentation

The afternoon began with a video presentation of three individual cases of vCJD with commentary from the neurologist involved, **Dr Holgar Alrogan**. Key points included: variability in the presentation of signs and symptoms; the different speed of deterioration from 12 to 27 months; the similarity of the late stages of the illness in each individual. The effectiveness of drugs for problems such as anxiety, agitation, spasticity and the use of sedation to assist in management were also considered.

Funding care packages

Carol Bell from the Department of Health discussed the opportunities for partnership between health and social services in financing community care assessments, regardless of the disease or condition involved.

Stronger partnership arrangements between health and social services would in future be underpinned by the use of pooled budgets. A pooled budget could be put together for one or several people, or for a service for a group of individuals. The suggestion for a pooled budget could come from anywhere. It would have clear aims and outcomes but there would also be flexibility in how it was applied. However, it was important to note that there would not be any new money: it was about using existing resources. Essential to the success of pooled budgets would be a co-ordinated approach to commissioning services and an integrated approach to delivering them. The most influential professionals involved in pooled budgets would be local authority councillors, health authority chief executives and the commissioners of services.

Chairman of the Network, Clive Evers (left) and co-ordinator, Gillian Turner, at the Younger Person with Dementia conference in November 1999

A social worker's experience of vCJD

Mark Crowe, a social worker from London Borough of Haringey, described his experiences as the key worker supporting a person with vCJD and their family.

Mark first came into contact with the patient when she was discharged from hospital to home with no care plan. She was doubly incontinent and being looked after by her relatives in the family living room. The form used to carry out the assessment was very inadequate because of the complexity of the patient's needs.

Six hours day care and night care were arranged by a multidisciplinary team involving eight different professionals. The family was desperate for larger accommodation and very angry. The younger children were under considerable stress and eventually received counselling. After three months, the family decided to go to the media about their plight and the case was reported on the front page of the Daily Mail. After five months and a damning health and safety report, a four-bedroom house was found and the family moved. After six months the patient fell into a coma and later died.

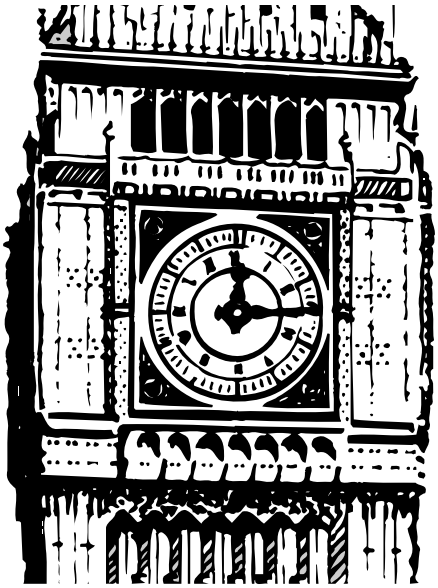
A carer's experience of vCJD

The last speaker was **Arthur Beyless** who spoke movingly and at length about his daughter Pam. She died from vCJD in 1997 after showing the first symptoms in the summer of 1996.

He highlighted a number of positive aspects of caring. Although Pam had stopped talking by April 1997 she was still able to communicate in different ways. Aromatherapy was introduced and she seemed to benefit from it greatly and appeared to respond to stimulation.

However, he regretted that Pam was not appointed a social worker when she was discharged from St Mary's Hospital and he didn't get a carers' assessment until he went to the media about the case. The physiotherapy that was eventually agreed came 14 months too late and it remains a mystery to him why high dependency funding was not made available to care for Pam.

CJD and the House of Commons



On 1 December 1999 a special adjournment debate took place in Parliament led by Tom Clarke, the Scottish MP for Coatbridge and Chryston. His interest arose because of the case of one of his constituents, Donna Marie McGivern, who died in 1999 at the age of 17. He also referred to the case of Steven McMurry in the constituency of Govan, Glasgow.

Mr Clarke outlined the problems faced by families caring for a relative with vCJD – financial difficulties, problems with transport, arranging childcare, and carrying out activities such as shopping and housework on very little sleep. He noted the enormous emotional strain on families and how they often have to argue vigorously for help and wait for professional assessments. He cited the difficulties experienced by Donna Marie's family in obtaining an appropriate wheelchair for her. By the time a suitable chair was found she was too ill to use it and confined to bed. Mr Clarke described the lack of a co-ordinated response to patients' needs and claimed that there was virtually no consistency of approach by local authorities or community health services in the UK.

He proposed the establishment of a central fund of £750,000 per annum for the sole use of vCJD patients and their carers. He asked what measures the Government had taken to support research into finding a cure for vCJD.

The Minster of State at the Department of Health, Mr John Denham, replied on behalf of the Government. He noted the need to develop a validated test for vCJD as recommended by SEAC and explained that the Department of Health is funding £3million of research into the disease. The Government was currently spending some £17million on research into this set of related diseases

He acknowledged that the management of vCJD needs to be rapid and flexible and reviewed frequently. A wide range of professionals would need to be involved. He referred to the crucial role of the National CJD Surveillance Unit in Edinburgh and claimed that NHS services for patients with vCJD were in place across the country.

Other rapidly deteriorating neurological conditions such as motor neurone disease also required a speedy and co-ordinated approach to health care. There are perhaps six patients suspected of having CJD and about 3,500 people with motor neurone disease. In that context, the Government had declined to establish a central fund specifically for vCJD.

Services need to be co-ordinated so that patients receive integrated care packages involving health and social care professions.Health and social services staff need to work together to ensure that their services are planned and delivered in partnership.

Provisions in the Health Act 1999 would allow the NHS and social services to overcome the barriers to joint working. For example, pooled budgets could be set up to provide responsive and flexible packages of care. Local managers would need to identify when that might be required, pool funds to provide a future care package, and agree in advance who should manage the pooled budget.

He drew attention to the backing giving by the Department of Health to the CJD Support Network for its case co-ordination initiative which aims to improve the co-ordination of care for patients affected by all forms of CJD. He also mentioned the support given to families concerned with vCJD through the Human BSE Foundation.

Occupational therapy with people with CJD

Ann Tweedie · occupational therapist

Ann Tweedie has been an occupational therapist (OT) for ten years and has specialised in clients with progressive neurological disorders for the last six. In the past 18 months she has worked with three people who have been suspected of having CJD. Here she describes issues realting to this emotionally demanding but rewarding job.

When I first meet a new client, whatever their condition, I explain my role. I then explore with the person and/or their family how the condition is affecting their daily life, often focusing on safety and independence in their home environment. The needs and wishes of carers are considered alongside those of the client.

If the person with CJD lives at home the OT has a significant role. We can advise on: how best to assist mobility (for example, how to help the person stand up from a chair); equipment to help improve safety (for example, bath equipment); methods to deal with confusion (for example, establishing a routine). I have found that agitation is the most difficult feature to manage in the home setting.

Issues that have arisen for me relating to clients with suspected or confirmed CJD include:

- Although I am a hospital-based OT, I am able to continue my involvement after the patient has left hospital. I suspect that this is unusual: most OTs work either in hospital or the community. Since I have known the client early in the development of the condition, I am able to assess changes in personality, behaviour or thinking. Also, if the client first gets to know me when their memory is relatively intact, trust is likely to continue. And I also believe that it helps the family to have someone 'with them' through the experience including post-bereavement follow-up. This continuity may be shared with the GP, social worker and, perhaps, the community nurse.

- A condition which changes so rapidly requires prompt, frequent and responsive input which considers future needs. This is very time-consuming and will often mean that the therapist may need assistance in covering the rest of their caseload. If the number of cases of CJD increases there will be an impact on the availability of OTs.

- The OT role overlaps with that of many other professionals (physiotherapy, speech therapy, mental health nursing, district nursing, social work). It can, therefore, meet some needs without the involvement of another professional. I knew my limitations, discussed queries with other relevant professionals and sought guidance on when to make a referral to a specialist. Limiting the number of professionals involved is a more sensitive approach. In some situations, the 'key worker' role is best carried out by a social worker.

- Working closely with other professionals has been very rewarding, in particular my relationship with a social work colleague and a number of district nurses. Communication has been excellent. The team has been a practical support to the client and family, and also a source of emotional support to me.

- The statutory, system for the provision of equipment is somewhat cumbersome, even on 'fast track'. I believe therapists should have rapid, easy access to a fund of money and/or a store of appropriate equipment to minimise delays (for example, specialist 'tilt in space' armchairs that cradle someone who has uncontrolled movements).

- The CJD Support Network has been an extremely helpful source of information and support. I am more confident in discussing issues with families and exploring the practical consequences of this devastating condition.

Contact an OT through the consultant, GP or local social services.

Letters

Auto immune response

Dear Editor
I write with a strong feeling of disappointment in response to 'BSE as an auto immune disease' by Professor Ebringer (CJDSN Newsletter, September 1999).

Professor Ebringer gained much publicity in May 1999 when he gained financial backing from the Ministry of Agriculture to develop this theory. To my knowledge and to date there have been no accredited published results regarding this 'theory'. I must profess an element of scepticism as the Ministry of Agriculture is facing costly litigation regarding their disastrous handling of the BSE situation.

All the most respected and eminent scientists in the field of CJD research associate themselves with the prion theory in relation to vCJD and BSE transmission. And I, as the husband of a vCJD victim, share the prion theory association.

When Professor Ebringer gained the funding, he made some highly insensitive remarks which caused upset amongst the families of people with vCJD. I hope, by publishing my letter, your readership can judge for themselves the true motives behind this theory.
Malcolm D Tibbert

Hidden diagnosis

Dear Editor
I sat down to read your newsletter yesterday and found myself crying my heart out for Mr Gerry Lyons. My father died from CJD six months ago. Like Mr Lyons' wife, no one seemed to know what was happening. The first indication something was wrong was when he telephoned me and said he'd forgotten how to write out a cheque. When the doctor asked him who the Queen was, he had no idea. It was obvious something was badly wrong. My father hated the idea of hospital. It took me three hours to persuade him to go, with the promise I'd get him home for the weekend. He never saw his home again. I will feel bad about that for the rest of my life.

He rapidly went downhill. From a man who trained as a tailor in retirement, making suits, curtains and sofa covers for all the neighbours, to a lost soul within weeks. His memory was so bad he forgot how to speak, then he lost all co-ordination. He lost the ability to walk, became incontinent and eventually couldn't swallow. During the last four days of his life he did not drink anything. From the phone call to the doctor to death it took six months.

Like Mr Lyons, I kept trying to find out what was wrong with my father. The hospital did test after test. They told me it wasn't Alzheimer's, it wasn't this, it wasn't that. They wouldn't tell me what it was. Eventually, a nurse told me off the record that my father's medical notes stated he had sporadic CJD. He said I wasn't to say who told me as he was worried he would get into trouble! So, like Mr Lyons, my father will never be a CJD statistic. It makes me very angry that the medical profession chose to keep this information from me. I will never forgive them.
Mandy Jones

Delayed diagnosis

Dear Editor
Mum fell and broke her left femur in February. She had a pin put into her leg the following day and, after some weeks' recuperation, she came home in the middle of March. As the only close relative, I looked after her at home for three weeks, but it slowly became clear that her mobility problem was not just due to the broken leg. Her co-ordination was not right and she was becoming uncharacteristically forgetful. I took her to her GP in April and the GP immediately had her admitted to hospital for tests. Mum was finally diagnosed with CJD in June and died in the early hours 15 days later.

As you well know, the disease has a completely devastating effect on the individual patient. It is devastating, too, for the relatives as they battle with their own confusion when confronted with the immensely disturbing effects of the disease. In Mum's case, a huge additional burden was created by the desperate pursuit of a proper diagnosis in the face of an almost total lack of interest from the medical staff. It was easier for them to put it down as a probable stroke, despite the lack of any hard evidence. I was upset and angry to find that a MRI scan had been ruled out – on grounds, I suspect, of cost combined with Mum's age of 72. She was, however, a remarkable youthful, vigorous and healthy 72 year-old still running her own business. Given Mum's robust general state of health, the hospital's lack of diagnostic action was scandalous.

Along with other relatives and friends, I could see the steep decline in Mum's mental and physical condition on a daily basis, but it proved next to impossible to get the doctors to recognise it. The consultant assigned to Mum went on holiday for a fortnight soon after she was admitted to hospital. It therefore fell to the house doctor to explain to us that, although she did not know the cause of Mum's condition, there was no hope of a recovery.

I could not accept this prognosis and referred the matter to the hospital chief executive to obtain a proper diagnosis. We succeeded – finally – in obtaining a second opinion from a brain specialist. The specialist really got things moving, immediately having Mum taken to his hospital and undertaking the tests I now know are for CJD.

Nothing could have prepared me for what he told us. The diagnosis of probable CJD, given on 1 June, almost two months after Mum had been admitted to hospital, was completely devastating. She died in hospital on 16 June and the cause has since been confirmed as CJD.

No one at the hospital was aware of the CJD Support Network. It was only by chance that a relative happened to see an article about the Network. At what was (and continues to be) the most distressing period of my life, I cannot say how much it meant to be able to speak to someone who knew not only the medical background but also what it was like, on a personal basis, to face this appalling situation. After so much lack of knowledge for so many weeks, we were finally no longer alone.
Paul Broadhurst

Editor: we welcome letters for publication. Please send to Gillian Turner (address on back page).

Infection control and the nursing care of patients with CJD

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Methods of preventing cross infection in hospitals has evolved from an understanding of micro-organisms and the effectiveness of various well-established decontamination methods.

Transmissible spongiform encephalopathies (TSEs) such as CJD pose a different set of challenges to the infection control team because the 'infectious agent' appears to be a pathological version of prion protein whose properties and routes of transmission are not yet fully understood. But it is recognised that the normal decontamination methods have had to be modified to reduce or destroy these abnormal prions.

The sorts of questions needed to address infection control matters include:

- How is the disease transmitted?
- What are the characteristics of the infective agent?
- Which body organs or substances pose most risk?
- Which people are likely to be most susceptible?
- How does one manage instruments and equipment?
- What is the risk to health care workers?

Regardless of the nature of an infective agent there are a number of universal precautions one can take in order to minimise the risk. These include:

- Handwashing between each patient contact
- Aseptic techniques when undertaking invasive tasks
- Maintenance of a clean environment
- Ensuring that effective procedures are in place for the cleaning and disinfection of reusable, multi-patient use equipment (for example thermometers, bedpans, stethoscopes)
- The wearing of personal protective clothing – for example, gloves, apron, visor – when undertaking tasks where one may be contaminated with high risk body fluids/tissues
- Incineration of all infected waste

In the case of prions additional precautions include:

- The use of single-use only equipment
- The pre-planning of all invasive activities and reduction of items used to a minimum. For example, blood taking and lumbar puncture
- Only allowing trained and experienced staff to undertake these activities
- Only allowing trained and experienced laboratory staff to handle highly infectious material. For example, brain tissue, spinal cord and eye tissue
- Limiting the number of people at brain biopsies to the minimum
- Undertaking such cases last on the list so that a longer time is available for cleaning to be carried out
- Following the guidelines on decontamination methods for prion diseases

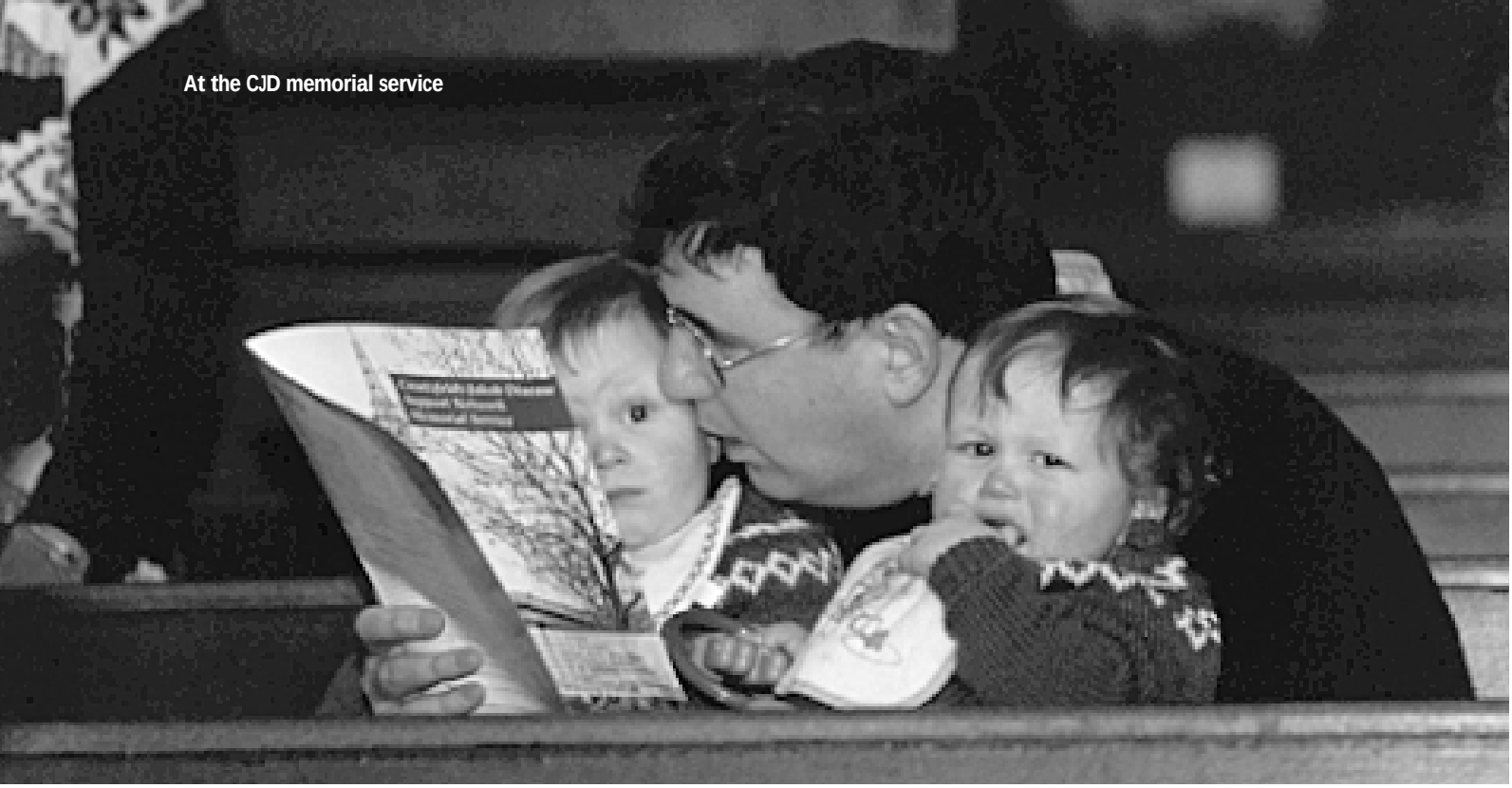
The bulk of nursing care for the prion disease patient requires the same standard of infection control as most other patients. They do not require to be placed in a side room for isolation. Managing day-to-day feeding and normal nursing activities pose no extra risk.

Further reading

For detailed advice, refer to:

Transmissible spongiform encephalopathy agents: safe working and the prevention of infection, the Advisory Committee on Dangerous Pathogens Spongiform Encephalopathy Advisory Committee, the Stationery Office, 1998.

Variant Creutzfeldt-Jakob disease (vCJD): minimising the risk of transmission. Health Service Circular HSC 1999/178, NHS Executive, 13 August 1999.



At the CJD memorial service

Alzheimer's Society membership application

he CJD Network is part of the Alzheimer's Society. ecoming a member of the Society adds to our strength nd 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 he Alzheimer's Society's monthly newsletter.

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

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ease make cheques payable to **Alzheimer's Society**

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

Name .....

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ostcode .....

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

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

CJDSN

CJD Statistics

Department of Health monthly figures of the numbers of deaths of definite and probable cases of CJD in the UK.

Year	Referrals	sporadic	iatrogenic	familial	GSS	vCJD	Total
1993	78	38	4	2	2	-	46
1994	116	51	1	4	3	-	59
1995	87	35	4	2	3	3	47
1996	134	40	4	2	4	10	60
1997	161	59	6	4	1	10	80
1998	152	61	3	3	1	17	85
1999*	159	48	4	0	0	11*	63

* Up to 31 December 1999, there have been 52 definite and probable cases of vCJD (including one case who died January 2000)

Source: DoH circular 2000/0071 (7 February 2000)

CJD SUPPORT NETWORK

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

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