CJD SUPPORT NETWORK

Helpline +44 (0)1630 673973

www.cjdsupport.net

Newsletter

Issue 23
May 2014

CREUTZFELDT-JAKOB DISEASE SUPPORT NETWORK NEWSLETTER

Parliamentary inquiry on blood, tissue and organ screening for vCJD

A parliamentary inquiry has been launched into the safety of blood, tissue and organ screening following fears that vCJD may be being spread by medical procedures.

Andrew Miller MP, Chair of the Parliamentary Committee said 'We want to explore whether the Government is taking this threat as seriously as it should be. Our new inquiry will investigate these issues in more detail and consider ways in which the UK can protect its vital supply of donated blood, tissues and organs.'

The Committee is exploring the national surveillance system for transfusion-transmitted infections and is considering the reporting of potential cases of CJD and the work of the National CJD Research and Surveillance Unit. The Committee is also discussing the development of technologies to reduce vCJD transmission risk via medical procedure.

This enquiry involves both written and oral evidence and the CJD Support Network was invited to make a written submission. What follows is a summary of the main points we made to the enquiry on 15 January 2014.

The submission

The submission opened with a brief summary of the Network's aims, work and membership, including its work supporting the two UK centres of excellence (the National CJD Research and Surveillance Unit in Edinburgh and National Prion Clinic in London). We outlined the scope and limitations of our response and explained that members with potential conflicts of interest had seen but had not contributed to our submission. Our submission continued under the following headings:

Scope of our response

The CJD support Network is only qualified to comment on issues related to communication with and support of patients, families and carers and with professionals. We have not addressed any of the science involved in the project.

Test for CJD

We welcome a valid and reliable test for all strains of CJD to further protect public health and to enable early diagnosis and access to CJD care.

Public health

Until a test is in place all current precautions to protect public health should be fully applied and monitored with utmost diligence.

Costs and funding

Although appreciating that costs will always be an issue and will be balanced against current and future benefits, we ask that costs are not the limiting factor in the decision making processes.

Communication protocol

We would urge that future communication, support and counselling (by specialist counsellors who fully understand the elements of this complex disease) are comprehensively addressed in a consistent, structured and co-ordinated

Continued on page 11...

Running for the CJD Support Network

Elliot Nobel took part in the London Charity 10K to raise money for the CJD Support Network. Thank you Elliot!

More fundraising news inside...

CJD SUPPORT NETWORK

We only have space to mention a few names in this newsletter but we value all our wonderful fundraisers. Even the smallest amount raised can help us to support a family affected by any strain of Creutzfeldt Jakob Disease.

Below are a few examples of the fundraising events that have been organised on our behalf in the last year.

Fundraising – a piece of cake!

Lucy Roberts

I lost my dad, Julian, to sCJD four years ago. Since he died we as a family have tried to think up lots of different ways to raise money for the Support Network. As it tends to be the same friends we approach for fundraising, it's nice to have an event where everyone benefits in some way, especially in a recession! A clothes swap is a really easy and fun way to make a bit of money, I held one recently and would really recommend it to anyone wanting to organize a charity event.

Firstly I decided on a date and then gave my friends plenty of warning so each of them could gather together enough to bring along. I asked them to put aside at least five items of unwanted but good quality clothing – including shoes, jewelry, hats and accessories, I gave them the date and time, promising tea, coffee, crumpets and homemade cakes and asked them each to bring along a £10 donation. I also asked them to RSVP and told them they were welcome to bring a friend.

In the fortnight before the swap I made a few different cakes and put them in the freezer so I wasn't too busy and stressed on the day. I asked around my friends to see if anyone had a clothes rail I could borrow and someone did! We set it up in one room downstairs and decided to use to the two bedrooms upstairs as the trying on rooms.

That afternoon we had a houseful of lovely people, all drinking, eating and chatting together. We waited until everyone had arrived and then we all crammed into the clothes room picking out things that took our fancy. Everyone went away with a few new garments, and got rid of clothes they didn't use anymore. One friend who couldn't come even sent her partner round with a tenner, a bag of clothes and the instructions to find her a new scarf!

It was a great success, and for very little effort we raised over £100 in the space of two hours just trying on clothes and eating cake!

Brenda Slater, age 62, ran in the London Marathon to raise money for the CJD Support Network in memory of her husband Brian – well done Brenda

Lower Morden Lane Lights Charity

Each year the residents of Lower Morden Lane decorate their houses with Christmas lights for charity. These attract a lot of attention and give much pleasure to many car loads of children. Each resident can choose their own charity and Sylvia Queensborough chooses the CJD Support Network.

Tough Mudder Charity Race

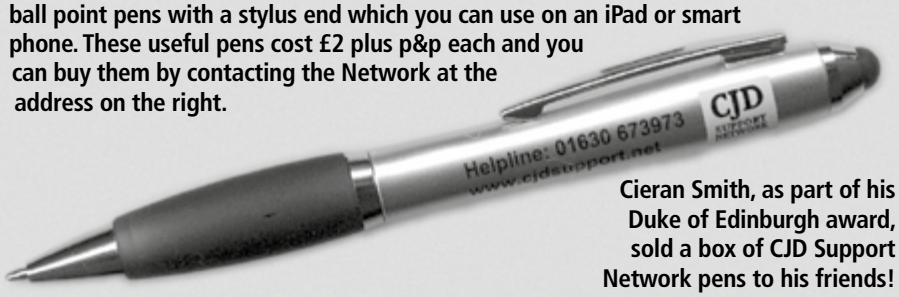
Richard Tittensor together with a team of friends entered the 2013 Tough Mudder Charity Race to raise money in memory of his dad Alan. I do not know whether you have seen this race but Richard said after he had completed the course, "it was so enjoyable and to be honest a privilege, although after two showers and a 30 minute bath, I still had grey towels! We're doing another in November so hold tight..."

Open garden

Robert Hirschhorn and John Hall open their delightful garden every year as part of the Camberwell Grove Gardens scheme in London and donate part of the money raised to the CJD Support Network.

Pen with stylus for iPad or phone

To raise money for the network we are selling CJD Support Network branded ball point pens with a stylus end which you can use on an iPad or smart phone. These useful pens cost £2 plus p&p each and you can buy them by contacting the Network at the address on the right.



Cieran Smith, as part of his Duke of Edinburgh award, sold a box of CJD Support Network pens to his friends!

Donations in memory

Heartfelt thanks to the families and friends of those below for donations received in their memory between March 2013 – April 2014.

Julian Bailey
Patricia Anne Barnes
Peter Barnett
Alan Bedford
Gillian Christine Bonser
Brenda Boseley
Helen Bullock
Valerie Chadwick
Janet Elizabeth Clarkson
Brid Collins
Ian Cox
John Cutting
Linda Fanning
Terrence Flatt
Darryl Freeman
Mick Golding
Rosemary Grace Harrison
Jesse Hobby
Jeanne Hooper
Marie Hutchinson
Iris Helen Inkpen
Beryl James
Brian Kaye
Tony King
Roger Lovell
Jacqueline (Anne) Morris
Anne Morris
Jean Lesley O'Brian
Vicky Parsons
Margaret Sedgwick
Brian Slater
Joanne Smith
Kenneth Trew
Jean Waterman
Pauline Whorlow
Linda Young

For any fundraising news of your own, or to make a donation, please contact: Gillian Turner, CJD Support Network, PO Box 346, Market Drayton TF9 4WN
Telephone 01630 673973
Email gturner@cjdsupport.net
Website www.cjdsupport.net

CJD figures

Recent figures from the National CJD Research and Surveillance Unit in Edinburgh

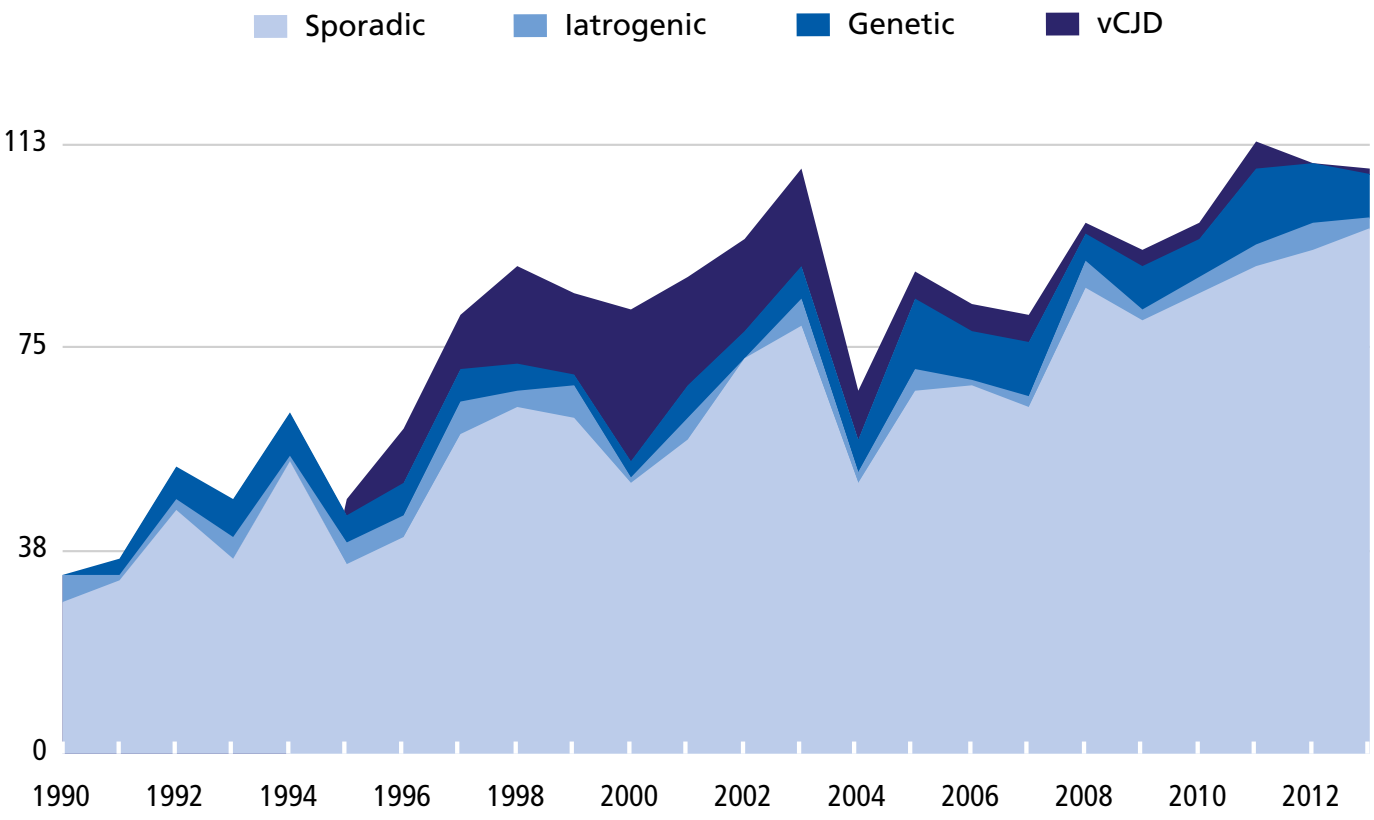
Recent numbers of deaths of definite and probable cases in the UK

Year	Sporadic	Iatrogenic	Genetic‡	vCJD	Total
2010	85	3	7	3	98
2011	90	4	14	5	113
2012	93	5	11	0	109
2013	97	2	8	1	108
2014*	20	0	1	0	21

*As at 7 April 2014 ‡Includes all genetic prion disease, including GSS

Definite or probable vCJD cases (dead and alive) in UK to date	176

Recorded deaths for the years 1990–2013



Introduction to prion diseases

Dr Louise RR Davidson, Clinical Research Registrar, National CJD Research and Surveillance Unit Edinburgh

Back to basics

Prion diseases are easier to understand if we firstly have a basic understanding of some key concepts. This includes the brain, proteins and genes.

The brain and cells

Everything in our bodies from our hair to our skin and organs are made up cells. Different types of cells exist and thereby allow the different parts of our body to function as they do. For example, our brains are made up of special cells called neurones. Neurones are electrically active and communicate with each other by sending electrical messages through junctions called synapses. It is estimated that our brains are home to around 100 billion neurones and over 100 trillion synapses which form a very sophisticated and intricate messaging system within the brain and allow us to perform complex brain function such as moving, thinking, emotion and sensation. Some of the functions of our brain is localised to specific areas. For example, memory and language is organised at the side of the brain and vision at the back of the brain. Therefore, if disease affects the brain, the anatomical area affected by that disease will dictate what symptoms we have or what function is impaired.

Proteins and genes

There are approximately 50,000 different proteins in our bodies. They are vital structural and functional components of the body and are made up of a string of building blocks called amino acids. Each protein and its string of amino acids fold into a complex shape. It is the specific shape that a protein folds into that is key to its function (i.e. if a protein loses its shape then it ceases to work normally).

All of our proteins are made in cells. Each cell contains a nucleus and within each nucleus are our chromosomes. We receive a set of chromosomes from our mother and another set from our father. Chromosomes are long chains of DNA and a tiny section of DNA is called a gene. Each gene is a set of codes. These codes provide the instructions that make our string of amino acids and hence our proteins.

If our genes have certain variations or errors in them, then this subsequently causes a change in the protein that it is responsible for making. Errors in the gene code can lead to a change in the string of the amino acids, a change in the protein and its shape and therefore its overall function. This chain of events can subsequently cause disease. These errors are referred to as pathogenic mutations.

Interestingly, there can be variations (i.e. not errors or mutations) in our gene codes that are not directly disease-causing although can have an indirect importance which will be discussed later. These variations in gene code are termed polymorphisms and are found distributed in the normal population.

What is CJD and prion disease?

Prion diseases or 'transmissible spongiform encephalopathies' are a set of neurodegenerative brain diseases that affect both humans and animals. Prion disease is essentially a single disease taking different forms in humans and animals.

Unfortunately, these diseases are progressive and remain untreatable. The hallmark of the disease is related to an abnormal folding of a specific protein, the prion protein. The normal prion protein is found in many body tissues but there is a huge concentration of them in our brains. Their exact function remains elusive, but we need them for normal brain function. When we discuss the prion protein, we often refer to the normal prion protein as PrP^{C} and its abnormal, misfolded form as PrP^{Sc}. When the abnormal protein is assessed in the laboratory, there are two recognised patterns of misfolding that can occur and these have been classified as Type 1 and Type 2. The precise mechanism of how the misfolded prion protein leads to disease in the brain remains unclear.

We have discussed that different genes code for different types of protein. The human gene that codes for the prion protein is located on chromosome 20 and is called the prion protein gene or PRNP for short. Pathogenic mutations in this gene are responsible for genetic prion diseases. However, as we previously touched upon, not all variations in the genetic code lead to disease (i.e. polymorphisms). In PRNP there is a known polymorphism or code variation at code position 129. At this position on the prion protein gene, the code can make 1 of 2 amino acids, Methionine (M) or Valine (V). As we inherit a set of chromosomes from both our mother and our father, we receive two sets of PRNP.

Therefore, each and every one of us are either codon 129 MM, MV or VV depending on what code variation we received from our parents.

What is important to remember is that these polymorphisms are not disease causing BUT do have the ability to influence disease. For example, they can modify the risk of getting a prion disease or affect the clinical picture of the disease. In the UK, approximately 50% of the population are MV, 11% VV and the remaining 39% MM.

Human prion disease

The commonest form of human prion disease is Creutzfeldt-Jakob Disease (CJD). Human prion diseases remain progressive, 100% fatal and are rare (1-2 deaths/ million/year). They are broadly classified into three categories: **idiopathic; acquired; genetic**.

Idiopathic human prion disease

The term 'idiopathic' essentially means of unknown cause and in this context refers to Sporadic CJD (sCJD). This form of CJD typically targets the middle-age and elderly age group. Although the cause remains unknown, individuals who are codon 129 MM are more susceptible to the condition. In contrast, those who are codon 129 MV are less susceptible.

Sporadic CJD can affect the brain uniformly or alternatively can be somewhat 'patchy'. The classic clinical picture typically starts with a rapidly progressive illness involving dementia (confusion, memory problems, language problems), ataxia (incoordination and unsteady gait), visual disturbance and myoclonus (jumpy, jerky movements). The average prognosis from the initial symptom is usually under 6 months and frequently only around 4 months.

Approximately 20-25% of individuals present somewhat atypically. In such circumstances, the age of onset is often younger and has a slower progression of illness. In addition, they can present with isolated focal symptoms (e.g. ataxia or visual disturbance) for a significant period of time before going on to develop the more classic clinical picture. To some extent, the differences in clinical presentation are associated with the neuropathological findings, the codon 129 polymorphism and the different misfolding conformations of the abnormal prion protein PrP^{Sc} (i.e. type 1 or Type 2). Therefore, sporadic CJD is broadly classified into six subtypes: MM1; MM2; MV1; MV2; VV1; VV2.

Acquired human prion disease

Iatrogenic (iCJD)

This refers to CJD that has been acquired through medical/surgical procedures or treatments. The most common instances relate to the use of cadaveric derived human growth hormone (used to treat short stature) and human dura mater (fibrous membrane that lines the brain and spinal cord and used in neurosurgery). Iatrogenic CJD can present at any age and has a clinical presentation similar to that of sporadic CJD. However, in cases associated with growth hormone treatment, the clinical presentation is often an isolated ataxia.

Variant (vCJD)

This refers to CJD contracted through the ingestion of BSE (Bovine Spongiform Encephalopathy) contaminated food from cattle. Dietary protection measures were implemented in 1989/1996. To date, there have been no new cases of vCJD in an individual born after 1989.

In contrast to sCJD, vCJD presents at a considerably younger age with an average age of onset of 28 years old. The illness also presents differently with early psychiatric/ behavioural symptoms and painful limbs. The test results are also different which will be discussed later. In addition, the duration of illness is slightly longer and is around 14 months. Interestingly, all cases of definite or probable vCJD (as defined by WHO diagnostic criteria) were codon 129 MM.

Genetic prion disease

Genetic prion disease results from a pathogenic mutation of the prion protein gene, PRNP. There are many different types of mutation and these are dominantly inherited through the generations (autosomal dominant). This means that 50% of the offspring of a carrier of the mutation are affected by the condition. Women are affected just as much as men. Clinical presentation is variable and partly depends on which type of pathogenic mutation is responsible. There are some forms of genetic CJD that can mimic the presentation of the sporadic form. There is usually a strong family history of the condition, but not always, and therefore the only way of absolutely excluding a genetic form of CJD is through blood testing.

How is CJD diagnosed?

CJD can only be absolutely diagnosed by neuropathological examination of brain tissue. This is achieved following post mortem examination or more rarely, by brain biopsy. However, we can diagnose it in life with high reliability and confidence through a suggestive clinical picture, exclusion of other possible diagnoses and supportive investigations.

The investigations that we find useful when diagnosing CJD include the electroencephalogram (EEG), MRI (magnetic resonance imaging) and testing for a protein marker called 14-3-3 in the spinal fluid. Although these tests can show changes that support CJD as a diagnosis, they are not absolutely specific for the condition and can be found in other illnesses, which is why they cannot absolutely diagnose CJD. Recent developments are promising with a blood test for vCJD and a new spinal fluid test (called RT–QuIC) for sporadic CJD. Tonsil biopsies can also be very helpful in the diagnosis of vCJD.

As we have discussed earlier, genetic forms of CJD can be diagnosed by looking for mutations in tissues such as blood. Iatrogenic CJD is highly suggestive if the appropriate clinical picture is associated with previous exposure to a known risk factor.

My experience attending the Family Support Meeting of the CJD Support Network

Dr Louise RR Davidson
National CJD Research and Surveillance Unit, Edinburgh

Two and half years ago, I stepped out of my registrar training in neurology to join the Edinburgh CJD Research and Surveillance Unit as their clinical research registrar. Prior to that, I had limited experience in CJD but was very keen to learn more about this devastating illness. As many of you know, the role of the research registrar is to meet with patients and families. As well as assisting with the diagnosis and asking questions to help with disease surveillance, I personally feel that one of my most important roles is to ensure that this complex disease is explained in a way that can be understood and that patients and families are well supported.

I recognise that it can sometimes be difficult to take in the vast amount of information that we give during our visits as it is clearly an extremely difficult and emotional time. As such, it was invaluable to be given the opportunity to speak at the CJD Support Network meeting in Birmingham and may I thank the Support Network organisers for inviting me. It gave me the chance to explain the disease in more detail and demonstrate how important and helpful the information we receive from relatives is to learning and understanding more about this condition.

We are very grateful to all of the families who meet with us and for answering all of our questions. I always reflect upon each meeting I have and often wonder how well I explained things. As doctors, we routinely speak using medical jargon and it is easy to forget that when we are trying to explain complex conditions such as CJD, it is extremely important to relay it in way that can be widely understood and in a non-patronising, sensitive manner. During my time working for the CJD unit, I have learnt a great deal about how communication between doctor, patient and their family is of paramount importance and this has made me think about my clinical practice in general and how I can improve as a doctor.

In medicine, it is common practice to attend and present at educational meetings and these can be at departmental, regional, national or international level. I have given a number of presentations over the years but I will be honest and say that I have never been more nervous than I was when I spoke at the meeting in Birmingham. The main reason for this was the fact that my audience were people who have been directly affected by what we all know is a terrible illness, and I was about to discuss it in detail. What I found invaluable though was listening to the questions and discussions after the presentations as it highlighted to me where the main body of frustration lay – providing an earlier diagnosis and, of course, treatment. As discussed at the meeting, our unit in Edinburgh is working on a new spinal fluid test called RT-QuIC which will hopefully help with the diagnosis of sporadic CJD.

During the course of the day, I had an opportunity to speak to many families, some of whom I had already met in a professional capacity. On a personal level, I was humbled by the strength of character, fortitude and family unity that was displayed and this is something that I will always remember. It was a pleasure to meet you all.

Our next Family Support Meeting

The 2014 Family Support Meeting is on Saturday 15 November from 9.30–3.30 at the Burlington Hotel, Burlington Arcade, 128 New Street, Birmingham B2 4JQ. We do hope you will be able to join us, as so many families who have attended say how helpful and informative these days are and it is a great opportunity to meet and talk to other families who have had a similar CJD experience.

Details of the meeting will be sent out nearer the date but if you have any questions, please do not hesitate to call Gillian Turner on 01630673973

My husband Terry Flatt

by his wife Jackie

My husband recently passed away after having been diagnosed with sporadic CJD. I know my daughters have spoken with you and also friends have rung you for more information but this is the first time I have been in touch. I wanted to tell you Terry's story so that it may help others in dealing with this horrific disease.

Terry was a healthy 54 year old. He worked as an electrician and was mad about motor racing – his hobby was saloon car racing which is something he did for many years.

On Monday 15th of April this year he asked me to make him a doctor's appointment. This was a shock for me as he never went to the doctors. When I asked him why, he said he had been losing weight and was concerned. I knew he was worried as he had lost both his mother and father to cancer. I made the appointment for the Friday and we carried on as normal. On the Wednesday he was at Mallory Park race circuit testing his car out for the new season. He went to the doctor on Friday 19th April and when he came home I asked what the doctor had said and he told me he had to have a blood test, which he couldn't do until Monday. I needed a lift to meet a friend that evening and as he was driving he told me he was finding it very hard to concentrate. He got me to where we were going and went home. This was the last time he drove a car.

The next morning the confusion seemed worse – he said his head seemed muddled. I decided that instead of waiting for the blood test on the Monday and the week for the results I would take him to A&E. We spent a few hours there and they decided he needed to be referred for a CT scan but our GP had to do that, so first thing Monday we were back at the doctor. He made a call to the Medical Assessment Unit and Terry was sent there. He underwent numerous tests, including a neurological test, CT scan of the head, X-rays and ultrasound. All the time they ignored the confusion. After four days of tests we were told he had a probable lymphoma of the stomach and were sent home.

The confusion was getting worse so I took him back to the hospital. We also noticed he was walking funny and his speech was getting very laboured. A different doctor arranged a lumber puncture and an MRI scan, both of which we were told were clear. We went home again but a few days later he was getting worse. So back we went again. This time they did an EEG. We went home again – this was over a period of two to three weeks. Still no improvement so we went back yet again. This time we saw the neurologist who first examined him and who was shocked at the deterioration. He organised another lumber puncture, which was sent to Edinburgh for testing and an EEG and he went back to look at the MRI again. On the 15th May he told us that he suspected sporadic CJD but needed a specialist to look at the MRI. Apparently when they first looked at the MRI they were looking for tumours so did not recognise anything else. After this he confirmed our fears that it was terminal and he said it was six months to a year life expectancy.

We were then passed on to the palliative care team and sent home on Friday 17th May. On the Monday a nurse from the Princess Alice Hospice in Esher came to visit us and to assess what help we needed. She decided the best thing was for Terry to go into the hospice so he could be properly assessed. This was on the 22nd May.

Terry never came home again.

By this time he couldn't speak very well and his mobility was deteriorating rapidly. He was unable to do simple things like brush his teeth and was very, very forgetful. Between myself and my daughters Sian and Becky, Terry's sister Pauline and his brother Keith who flew in from New Zealand, we stayed with him 24 hours a day. The hospice staff were amazing, their care was second to none and we could not have coped without them. A few days after he went in he was unable to feed himself or get to the bathroom, so he was totally reliant on all of us.

We were visited at the hospice by both the doctors from Edinburgh and the National Prion Clinic in London who were very helpful in giving us information about the disease.

We spent our days with him sitting in the garden, my daughter brought

our baby granddaughter up every day and he was still recognising people although he couldn't communicate and was very frustrated by this in the beginning. After a while it was as though he became resigned to the fact that he couldn't talk so he didn't bother trying anymore. He was gradually withdrawing from us, although his favourite nurse, Sue could still make him smile.

By the beginning of June he was bedridden and couldn't do anything. On the 8th June his breathing changed. It became very shallow and rapid and we were told it could be a matter of hours that he had left. However, as he was only 54 and was a fit healthy person, he stayed with us until the early hours of 13th June where he passed away peacefully in his sleep. We were all there with him, four of us sleeping on mattresses on the floor.

We were told no autopsy would be required because the doctor from Edinburgh was confident that it was sporadic CJD because of the symptoms and rapidity of it.

The reason I want people to know Terry's story is to show how rapidly this disease can affect someone. From the first sign of confusion, on the 19th April, to the diagnosis on 16th May, to his death on the 13th June was less than two months.

The One in a Million appeal raises £2,000 for CJD

One in a Million is a charity appeal run by Gingernut Creative to raise money for the CJD Support Network. It's a tribute to the one in a million people who get Creutzfeldt-Jakob disease.

The campaign was created in 2013 in memory of Roger Lovell. To raise money for the CJD Support Network that helped Roger and his family, Andy, Roger's son, and his colleagues at Gingernut Creative are challenging themselves with a host of million-busting missions by doing a bunch of daft and difficult challenges.

So far the team has skydived, held bake sales, organised football tournaments, run marathons, swum for miles, fasted and even cut the grass … with scissors! All their efforts, from the hare-brained to the lion-hearted, have raised more than £2,000 already – over two thirds of the way to their £3,000 goal.

The CJD Support Network relies on donations to keep their services open. They do a vital job helping people affected by Creutzfeldt-Jakob disease, but receive no government funding.

If you would like to support the CJD Support Network, please visit www.justgiving.com/teams/oneinamillion to donate to the One In A Million appeal.

One in a Million

Some of One in a Million's activities:

- Cycling 1 million metres in 5 days
- Cutting a million blade of grass with a pair of scissors
- Falling a million centimetres
- Charity cake sale
- 5 a-side football tournament

Reducing the risk of iatrogenic transmission of CJD by improving the cleaning of neurosurgical instruments

University of Glasgow: AJ Smith, T Tomkinson & DF Lappin

The emergence of vCJD in the UK has raised concerns over the possible risks of transmission of iatrogenic prion disease due to its resistance to inactivation processes such as, steam sterilization.

Of particular concern is the high levels of infectivity found in tissues classed as high risk for CJD (central nervous system and posterior orbit of eye) and the potential for residual infectious material to remain on instruments following cleaning and sterilization. There are currently no reported cases of vCJD due to cross-contamination of surgical instruments. However, risk assessments stress the importance of ensuring a high quality of current cleaning and sterilization processes. While the absolute risk of CJD transmission via contaminated surgical instruments is low, the potential risk has led to the development of a series of measures to limit transmission.

An unintended consequence of the move to larger Sterile Service Departments (SSDs) which are serving several theatre complexes in different hospitals is that there is an increase in the time prior to the cleaning and sterilization of surgical instruments. Several workers have demonstrated that as the drying time (>15 minutes) increases, instrument residues from theatre become more difficult to remove, increasing risks of iatrogenic diseases transmission. To overcome this problem, workers and Department of Health policy statements have suggested using either spray/foam agents or polythene bags to keep the instruments moist from point of use until cleaned.

Many of the studies that have investigated methods for retaining moisture around surgical instruments prior to decontamination have been undertaken on stainless steel tokens contaminated with artificial test soils and laboratory washing protocols.

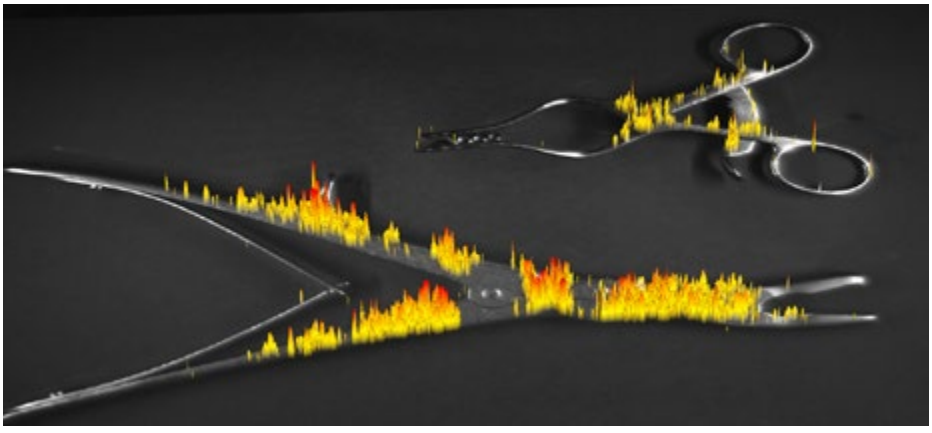


Figure 1: A ProReveal system 3D-fluorescence image of surgical instruments prior to the cleaning cycle. The residual protein is shown (yellow to red, which represents the relative protein concentration)

There is little published work that has investigated the application of instrument pre-treatment protocols under operating theatre and SSD conditions. Anecdotal reports have suggested that some pre-treatment chemicals may impair the function of the instrument washers or protect the soiling on the instruments from detergent action in the wash process.

Our group has obtained funding from the Scottish Infection Research Network (SIRN) to investigate a number of different pre-treatment methods to improve the cleanability of neurosurgical instruments. A number of different formulations will be screened in the laboratory and the two best performing agents will be trialed in the field with close collaboration with neurosurgeons, theatre staff, engineers and staff at the Cowlairs SSD. Improvements in cleaning will be monitored by measuring residual protein using biochemical assays contained in the British, European and International standard alongside a novel technique developed by Professor David Perrett and Dr Nanda Nayuni that visualizes residual protein on surgical instruments (see figure 1).

The success of this project is due to a collaborative approach between the following groups of organisations and people: University of Glasgow: AJ Smith, T Tomkinson & DF Lappin · NHS GG&C, Neurosurgery Dept: P Pamela & N Suttner · Cowlairs SSD: I McIvor, A Stewart, S McWililam & C Ladjadj · Health Facilities Scotland: S Holmes, R Allan, D Ingle, D Hill, P Howard, L Palmer, S Murphy & N McLean · Queen Mary University of London: D Perrett & NK Nayuni

National CJD Team

Margaret Leach and Blair Bathgate-Smith

Established by the Department of Health, the National CJD Care Team is based within the National CJD Research & Surveillance Unit and was formed in order to optimise the care of patients suffering from all forms of CJD. The national care coordinator post was established in February 2000 and in September 2001 the National CJD Care Team was formed. The present team consists of two care coordinators who are senior nurses with administrative and clinical neurological support from within the Unit.

Referrals and the family

When a referral is made to the NCJDRSU the research registrar will take that referral and, if appropriate, ask the care co-ordinator to attend that first visit to meet with the family. Once a diagnosis of probable or possible CJD is made, if the co-ordinator has not already met the family, the coordinator makes direct contact with them and offers the opportunity to meet and to assist with care planning. Once contact is made, the coordinator can meet on a regular basis with the patient, family and professionals involved in care. This will depend on need and will provide support and assist with coordination of local health and social care professionals. The coordinators provide valuable expertise in nursing patients with CJD and can anticipate and prevent some problems that may arise by offering skilled advice and education. The care team enables local teams to provide high standards of care and continues to be involved as long as needed. This does not always involve a visit in person. Contact by telephone, text or email is just as important and may be preferred by families and other professionals involved. Post bereavement support is offered to the family after the patient dies and assistance is given in accessing more specialised counselling.

CJD Care Package

The National CJD Care Team works in close liaison with the Department of Health and provides access to the CJD Care Package. This is a sum of money from The Department of Health which provides funding to assist local authorities with the care of patients suffering from all forms of CJD. The Care Fund is available to supplement local care and equipment provision. Health and Social Services will provide the basic elements of the individual patient's care package. The Care Package involves an individual assessment of need and will vary accordingly. It is essential that care packages are flexible and can change quickly to meet the rapidly changing needs of the patient. The aim is to provide a package of care that will meet the needs both for the patient and their family in a timely manner.

In addition to collaborations with national organisations in the United Kingdom, the Care Team liaises closely with international organisations, including the Australian and American CJD Support Groups and is an Official Friend of the CJD International Support Alliance.

The Care Team is available from 9am to 5pm Monday to Friday on 0131 537 3104. You can also email national.careteam@ed.ac.uk. Further information is available at www.cjd.ed.ac.uk

Continued from front cover

Parliamentary inquiry

way by all statutory agencies. This particularly applies to how, when, where and by whom a patient is informed of their heightened risk for the first time as well as their subsequent support.

Assuming those advised that they are 'at risk' are going to be in the thousands, a clear protocol, covering communication and support is required and must be understood and used by all agencies involved. We would ask that the CJD Support Network is involved in the development of such a protocol.

Discrimination and stigmatisation

We have current evidence that discrimination and stigmatisation are still occurring with those diagnosed 'with' or 'at risk' of CJD. It is therefore essential that protocols and information should also aim to eliminate the incidents of these in health and social care, as well as in other identified areas such as insurance, mortgages and the media.

Future funding

The CJD Support Network has no statutory funding and relies totally on public donations. An increased need for support for people who are informed that they are at risk of CJD would require additional funding for all agencies involved, both statutory and charitable.

Conclusion

The CJD Support Network's submission was acknowledged by the Committee.

All submissions have been published on the Parliamentary Inquiry on blood, tissue and organ screening for vCJD website at www.parliament.uk

CJD Support Network

Management Committee 2012

Professor Richard Knight, Chair Richard is a Consultant Neurologist at the National CJD Research and Surveillance Unit in Edinburgh

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

Mike Curtis, Treasurer
Mike is a former bank employee whose wife, Joyce, died of sporadic CJD in 2006

Sarah Tomkins
Sarah's late husband Edward died of sporadic CJD

Roger Tomkins
Roger's daughter Clare, died of vCJD

Alison Kenny
Alison's father died as a result of a contaminated blood transfusion. She is a RGN, nurse practitioner

Jean Bailey
Jean is a retired human biology lecturer. Jean's husband, Julian, died recently of sporadic CJD

Judy Kenny
Judy's husband, Deryck, was the first person to die of vCJD through a blood transfusion. Judy is a retired nurse

Derrick Biggs
Derrick is Head of Provider Services, Adult Social Care, and lead officer for Association of Directors of Social Services in matters pertaining to CJD

Dr Andrew Smith
Andrew is a Professor and Consultant Microbiologist at Glasgow Dental Hospital & School, University of Glasgow

Dr Simon Mead
Simon is a neurologist working at the National Prion Clinic

Sandra Walshe
Sandra is a Registered General Nurse whose sister in law died of Sporadic CJD

Gillian Turner
CJD Support Network co-ordinator

Can you help us this year to raise money?

Due to the present economic climate it is very difficult to attract grants, so fundraising by members and their families is even more necessary to maintain the work of the network. If you have any ideas or you would like help to arrange a fundraising activity, please contact Gillian Turner (see below).

The CJD Support Network was established in 1995 by relatives of people who have died with CJD and is now recognised as the leading charity for all forms of CJD. Our aims are:

- To offer support to individuals and families concerned with all forms of CJD.
- To offer support to people who have been told they are at a heightened risk of CJD through blood and surgical instruments
- To provide emotional support for carers and to link families with similar experiences of all forms of CJD..
- To offer small care grants for families in need whilst caring for a family member with CJD.
- To provide accurate, unbiased and up to-date information and advice about all forms of CJD.
- To provide a national helpline on all forms of CJD.
- To promote good quality care for people with all forms of CJD.
- To promote research into all forms of CJD and the dissemination of research findings.
- To develop a public response for all forms of CJD

Membership

Becoming a member of the CJD Support Network adds to our strength and enables you to take a full part in the decision-making process and the work of the Network. If you would like to become a member of the CJD Support Network and receive free regular copies of our newsletters and any other information we produce, please send £10 annual membership to the CJD Support Network, PO Box 346, Market Drayton, Shropshire TF9 4WN. Please make cheques payable to *CJD Support Network*. However, if you are caring for someone with CJD and would appreciate free membership, please tick this box ☐

Name ..... Title .....

Address .....

..... Postcode .....

Telephone ..... Email .....

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

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

Contact: Gillian Turner, CJD Support Network, PO Box 346, Market Drayton TF9 4WN
Telephone 01630 673973 · Email gturner@cjdsupport.net · Website www.cjdsupport.net
Charity number 1097173