

## New website goes live

**We are delighted to report that the CJD Support Network's newly commissioned website has gone live. We do hope you will take a look and find it interesting and informative.**

We have worked very closely with our web site developers, Extra Mile of Eccleshall. Our aim was to make the website, attractive to the eye, easy to navigate and for the information to be clear and easy to read. We have also developed the website suitable for mobile and tablet viewing as many of our families have told us that whilst they spent many hours at the hospital they often looked up information on the website.

We may have teething troubles in the early days and if you have any comments to make or suggestions for new sections please do not hesitate to contact me either by email or telephone.

We would like to thank you all who have made donations in memory of loved ones, fundraised by many different events or donated in any way. We are very grateful. The money we have received has enabled us to commission this new website which we hope will help new families to come in the future who search the internet for information and support.



The new website is much clearer and works well on mobile phones and tablets too

### CJD Support Network's free helpline number 0800 085352

In addition to our new website we have also been able to launch a free phone number on our helpline, thanks to your generous donations. The number is 0800 0853527. We do hope this will encourage callers to telephone us – whether they want to find out some information or they just want to talk to someone.

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# CJD Support Network invites you to the **Annual Family Support Meeting** Saturday 19 November 2016 · Birmingham

Our annual family support meeting will be on Saturday 19 November 2016. It will be held at the Burlington Hotel, New Street, Birmingham, starting at 9.30am and finishing at 3pm. If you wish to register for the meeting, please contact Gillian Turner at [gturner@cjdsupport.net](mailto:gturner@cjdsupport.net) or on our Freephone number 0800 0853527. There is no charge.

Attending the meeting is a great opportunity to meet and talk to other families with a similar experience of CJD. You will also meet world experts on CJD and be able to ask all those unanswered questions and listen to them sharing the latest information about this devastating disease CJD. We look forward to welcoming you.

## Evaluation of last year's Family Support Meeting

We thought you might like to read a summary of comments made by families who attended last year's family support meeting from our evaluation sheet. Those attending the meeting in 2015 overwhelmingly found the experience positive and the day beneficial.

### **What for you was the most beneficial part of the Family Support Meeting**

- Meeting new people and sharing experiences with others who are or have suffered with CJD themselves or a family member.
- Clearly given information by neurologists on a very scientific subject about the disease and treatment
- I was able to talk privately with a neurologist
- Making a less isolating experience of living with genetic CJD.
- I was alone and nowhere to turn. A magic phone call to Gillian Turner.
- Comforting to share experiences and medical info.
- Gave me hope having support from everyone.
- Because we had not spoken to anyone about this condition and felt quite isolated.

### **What was the least beneficial part of the Family Support Meeting for you?**

- Some people felt discussions about other strains was not relevant to them. Others felt this was nonetheless interesting
- One person felt the background story of the CJD Support Network was not relevant to them. However, another person said that they were interested to hear about the beginnings of the Network, as they had such respect and thanks for Gill and all who helped when life threw them this curved ball. [The background to the Network's beginnings was a one-off as part of last year's 20th anniversary – Ed]

### **Did the Family Support Meeting, in general, meet your expectations? If not, what expectation was not met?**

- Yes, I have attended several times and have always found the meeting to be very informative.
- We had no idea what to expect as it was our first visit but it was a much easier day than expected.
- Yes, it did and beyond.
- Very much so, but it was our first meeting, so few expectations. Nice to see so many leading professors talking positively.
- Yes, but maybe consider a location change?

This year's meeting is on Saturday 19 November from 9.30am to 3pm at the Burlington Hotel, New Street, Birmingham. To book places, please contact Gillian Turner on 0800 085 3527 or [GTurner@cjdsupport.net](mailto:GTurner@cjdsupport.net). There is no charge, but we need to be able to assess the number of attendees to plan the food and drinks.

# The Human BSE Foundation Memorial Service

Roger Tomkins

The Human BSE Foundation Memorial Service is held every year opposite the Houses of Parliament. It is always a very touching occasion. It started in 2000 after the publication of the BSE Report, which was prepared under the direction of Lord Phillips. Many families attended the BSE Inquiry and also gave evidence.

The Human BSE Foundation was formed in the early days of the emergence of vCJD. It followed the announcement in the House of Commons in 1996, by the then Minister of Health, that the disease which was Bovine Spongiform Encephalopathy (BSE) had been passed on to humans. This is now known as vCJD, a variant of the rare brain disease Sporadic CJD, or Classical CJD.

It was decided in 2000 that families who had lost loved ones to this terrible disease would meet every year on the



edge of the River Thames to remember collectively those who had died of vCJD. The Human BSE Foundation was given permission by the local authority to produce and erect a permanent memorial plaque on the wall of St Thomas's Hospital, exactly opposite The Houses of Parliament.

The memorial service is held every year on the nearest Saturday to the 16 March. This is the date that the then government admitted in the House of Commons that indeed vCJD was as a direct result of humans eating BSE

contaminated meat. It was therefore also felt to be an appropriate place for the annual memorial to take place.

At the memorial service the names or reference to every person who had died from vCJD are read out. It is not a religious service as there are many different religious faiths present, but being together on this one day every year means a lot to families who are still grieving after so many years.

A single white rose is placed in the River Thames for every person who has died, and it is a very moving moment watching all the roses quietly float down river. A wreath of yellow chrysanthemums is also placed underneath the memorial plaque.

Some of the families stay on to have lunch together, to talk about their lives as they are now.

*Roger Tomkins' daughter Clare died of vCJD on 22 April 1998 at the age of 24*

## The vCJD Trust Compensation Scheme

**In October 2000, The Secretary of State announced that the Government would pay compensation to the victims of variant Creutzfeldt Jakob Disease (vCJD) and their families. This fund is not available for any other types of CJD.**

### Compensation

The Trustees recognise that money cannot adequately compensate for the loss of a loved one to vCJD, but it is hoped that it will go some way towards reflecting the trauma and tragedy suffered by victims and their

families. Payments made whilst the victim is still alive may also go some way towards alleviating their suffering.

### The compensation fund

Consultations with representatives of families affected by vCJD were held, and details of the Scheme were announced on 1 October 2001. The Government has committed the sum of £67.5 million for up to the first 250 cases. The fund is divided into the Main Fund, for which £59.5 million is now allocated, and the Discretionary Fund, for which £8 million is now allocated. The total number of cases of vCJD is uncertain and the Government will review the Scheme if the total exceeds 250.

### Procedure

In early 2010 the Department of Health agreed to a revision of the Scheme in order to simplify the procedure

for making payments to Victims and their families and a revised Trust Deed was signed on 12 February 2010. The revised Trust Deed contains two versions of the Scheme, the first applies to victims diagnosed on or before 30 March 2010 and the second applies to victims diagnosed on or after 31 March 2010.

### Enquiries

All enquiries to the Trustees should be made to Fieldfisher, Solicitors, and directed for the attention of Jonathan Zimmern or Rose-Anna Lidiard on telephone number 0207 861 4000. The Trustees encourage families who want to make a claim for compensation to contact them direct, via Fieldfisher.

You can find out more, and make contact, through the website [www.vcjtrust.co.uk](http://www.vcjtrust.co.uk)

# How has the diagnosis of CJD improved?

**Professor Richard Knight outlines progress made in diagnosing CJD over the last thirty years**



Professor Richard Knight,  
Consultant Neurologist and Director  
of the National CJD Research &  
Surveillance Unit

**When things change, the changes are not always immediately obvious, especially when they occur step by step or gradually. My first involvement with CJD research was in the early 1980s. There are still difficulties in the reliable diagnosis of CJD but a look back over these 30+ years makes it clear that a lot of progress has been made.**

## Stages in diagnosis

Diagnosis of any disease relies on three broad steps: thinking of it, excluding other possibilities and having confirmatory test results.

In the case of CJD, the absolutely definite confirmatory test requires neuropathological examination of the brain and there are two ways to achieve this: Autopsy after death and brain biopsy in life. Biopsy is obviously a big step with some possible risks; it is undertaken rarely and arguably its main current role is in the exclusion or confirmation of illnesses other than CJD that could be treatable. Autopsy is obviously of no use to the living patient. Genetic testing, which can be done on a simple blood sample, is also possible in the case of genetic CJD.

Here, I will be considering diagnosis in life, of non-genetic CJD, without the use of brain biopsy.

## Early developments

In the 1970s and early 80s, clinical diagnosis was potentially problematic at all three steps. CJD was a rare illness, not familiar even to many neurologists, with incomplete knowledge of its clinical features; just thinking of it was not necessarily easy. The methods of investigating brain disease were relatively limited and, therefore, the consideration of alternative diagnoses was not always simple. Finally, the

only confirmatory test was the EEG. EEG recording involves placing small electrodes on the scalp to detect the electrical activity of the underlying brain. In many, but not all, cases of CJD, the normal activity is gradually lost and replaced by a rather characteristic abnormal pattern. However, aside from the fact that it is not always found, there were difficulties in defining exactly what should be taken as a truly characteristic pattern and, in any case, this pattern could be seen in clinical situations other than CJD.

Following a number of studies, including a five-year study in England and Wales, the clinical picture of CJD was better defined, the disease was more familiar to neurologists and there were significant developments in the investigation of brain disease (not least the development of clinical MR-magnetic resonance-scanning). Additionally, with the advent of BSE and variant CJD, in the 90s, CJD became very widely known both to doctors and the public.

## Improvements

From the 1990s onwards, the diagnosis of CJD was significantly improved: the diagnosis was more often considered (with good clinical guides as to when it should be considered), clinical neurological investigation in general was technically better and there were three potentially confirmatory clinical tests available: the EEG, the MRI and CSF protein tests.

The EEG remained in use but gradually took on a less important role as the other two tests moved into practice. Brain MRI has a double role of course: it is undertaken to exclude other possible diseases as well as to detect certain changes characteristic of CJD. Helpfully, there are different MRI changes that are characteristic of sporadic and

# It is likely that better outcomes will result from earlier, rather than later, interventions and this requires earlier diagnosis

variant CJD. CSF (cerebrospinal fluid) is obtained by performing a lumbar puncture (a needle is placed into the lower spine under local anaesthetic) and this too has a double role: the exclusion of certain other diseases and also protein testing to support a diagnosis of CJD. The original CSF tests looked for increased levels of normal brain proteins (the main two being proteins called 14-3-3 and S100b): in certain situations, elevated levels of these proteins provide strong support for a diagnosis of CJD. The combination of better disease recognition and these three tests allowed more straightforward and more reliable diagnosis of CJD. However, problems remained. Firstly, it was increasingly recognised that clinically atypical CJD cases occurred. Secondly, there were cases in which these three very helpful tests were negative. Finally, all three of these tests are essentially non-specific: they are not tied to the fundamental disease-process in CJD and the abnormalities characteristic of CJD can be seen in other clinical situations or diseases. Clinicians have to use these tests carefully and interpret the results in the overall clinical context. In fact, clinicians should generally always view test results in their particular context but tests which are incidentally indicative of a disease rather than a direct product of the disease-process are always less reliable. For example, several things may suggest that a patient's illness is due to HIV infection but a positive HIV test is a definitive proof of such infection.

## The CJD disease-process

So, what is the essential disease-process in CJD? The underlying molecular event is the conversion of a protein (prion protein) in its normal form (PrPC) to an abnormal form (PrPSc) and the deposition of this abnormal form in tissues (especially the brain). More

specific diagnostic tests depend on this molecular process.

## Developments in techniques

In this there are three important scientific developments: the understanding of the role of prion protein in CJD, the ability to detect the abnormal form (PrPSc) and the invention of techniques to amplify small amounts of PrPSc. This last development is important as the amount of PrPSc present in certain situations may be too small to detect by our otherwise effective standard methods. The amplification techniques essentially rely on what was discovered about the prion protein process in disease: once PrPSc is present in the brain, it can interact with the normal PrPC causing it to convert into more PrPSc. This is one important aspect of the disease development and progression in affected brains. There are two main amplification methods: PMCA and RT-QuIC. While they both essentially try to mirror the way PrPSc causes PrPC to form more PrPSc, there are important technical differences between the two techniques, and also variations on these methods, that we can leave aside in this article.

## Disease-process specific tests

The disease-process specific tests are as follows:

- 1 The specific test that is presently a routine part of investigation is the CSF RT-QuIC test for sporadic CJD. This is a very sensitive and highly specific test for this form of CJD.
- 2 Another test is the Direct Detection Assay blood test, developed by the MRC Prion Unit in London, that is relatively sensitive and highly specific for variant CJD.
- 3 A very recent study has reported a test based on urine in variant CJD (using the PMCA method).

- 4 Another interesting test possibility is one based on nasal brushing or swabbing. Inside the top of the nose, there are specialised cells for the sense of smell. These are actually specialised neurones that are effectively outgrowths of the brain, but which are easily accessible through the nose. Because of their neural nature, they may contain PrPSc in CJD and nasal samples can be tested for the abnormal protein, using amplification techniques such as RT-QuIC if necessary.

It is interesting, and puzzling, why these tests, all based on detecting abnormal PrPSc, can behave differently in sporadic and variant CJD.

## Summary

In summary, there has been a lot of progress in diagnosis with, in recent times, the development of highly specific tests based on the basic molecular disease process underpinning CJD. One might ask why such progress has taken so many years, but, as indicated above, there are several steps involved and one should not underestimate the time it takes to make scientific advances, to develop reliable relevant laboratory techniques and then to assess any potential tests in real life practice. These advances represent a great deal of time, effort and money.

Naturally, diagnosis has importance in allowing understanding of what is happening to people affected by illness and in planning their care. However, effective treatment for CJD still eludes us. As treatments are suggested and developed, it is likely that better outcomes will result from earlier, rather than later, interventions and this requires earlier diagnosis. These modern, disease-process specific tests should aid earlier diagnosis.

# The role of the CJD Research and Resource Centre at the National Institute for Biological Standards and Control (NIBSC)

**Dr Philip Minor give an overview of the role NIBSC has in assessing possible diagnostic tests for CJD and in collecting scarce patient samples to help in the development of tests**



Dr Philip Minor, Head of Virology Division, National Institute for Biological Standards and Control

## The need for a test for CJD

There is still a need for a reliable test to identify individuals incubating CJD and vCJD in particular. Those who have been informed that they are at higher risk of infection because of medical or other treatments may wish to know whether they are really infected or not; from a public health point of view it would be helpful to identify people who are potentially infectious for others, so that spread for example through blood transfusion could be prevented. Complicated precautions and models have been developed to minimise possible onward spread and to predict future incidence of disease. All are based on worst case assumptions including estimates of the incidence of currently silent infection which have been hard to come by and are not that certain.

## The problems in developing a test

Development of a test for any infection is always fraught; a test that incorrectly misses an infected sample or incorrectly identifies a normal sample as infected has major bad effects for the patient and the public health system. Tests for CJD are even more difficult than most because the infected state is almost entirely without symptoms or any marker of infection before symptoms develop. Virus infections are associated with virus specific nucleic acids or proteins or antibodies made by the patient against the virus and assays for infection can be based on these associations which do not exist for CJD. In addition the number of cases is thankfully smaller

than for most other infections; it is not hard to get a thousand specimens for developing a test for Hepatitis B for instance which would be unthinkable for vCJD. Moreover vCJD is a stealthy and frightening disease, so in the early days at least the desire for a test was very great, which created the possibility of poor tests being developed and used. There needed to be some kind of objective scientific evaluation and judgement of tests in development, to see if they were likely to give reasonable results when tested with the scarce real patient samples.

## NIBSC

The Department of Health therefore funded NIBSC to establish a collection of samples relevant to test development and devise a route by which promising tests could get access to the few real samples from patients that were available. The centre provides a clear if torturous route for developers to do this and the Oversight Committee considers proposals for studies and guides its activities.

## The Oversight Committee

The Oversight Committee was set up to include all groups that might have a technical interest or ability in CJD test development. It is intended to access the best possible relevant scientific advice and includes CJD scientists and medical experts from the MRC Prion Unit and Edinburgh Research and Surveillance Centre, as well as scientists from the agricultural sector with an interest in BSE or scrapie and possible diagnostics being developed there. Public Health England (PHE) and the blood services are included.

NIBSC organises the meetings and brings proposals and results to the committee for discussion. Developers are identified through scientific contacts and meetings and are usually contacted by NIBSC scientists, but things can be brought into the system through other committees such as the Prion Working Group (PWG) of the blood transfusion service or the TSE working group of the Advisory Committee on Dangerous Pathogens (ACDP). The idea is to be as inclusive as possible. Most of the meetings are face to face unless an urgent response is needed and a meeting proves difficult to organise.

Developers who wish to access samples through the committee follow an established route, with the results being discussed at each stage by the committee members, to see if it is good enough to continue to the next step. It is up to the developer to decide whether they wish to continue; they may fail at a step, but if they wish to modify the assay and have another go that is welcomed.

### **Blood test**

Blood is the easiest sample to get for testing and the amount of infectivity in blood is known to be low. The initial evaluation involves measuring the sensitivity of the test by trying it out on samples that are known to be infectious, such as dilutions of infected brain. Blood samples from healthy donors are then tested to establish whether the test incorrectly scores samples positive.

This has been followed by the use of samples from animal models which are easier to obtain; assuming the test will work on animal samples, this can show that detecting signals is possible. It is also possible to look at animal samples in the preclinical stage, which have been shown to be infectious, for example by transfusion to recipient animals. If a test cannot detect such samples it is obviously not much good in practice.

The experiments needed to collect these kinds of materials take a very long time to complete and are very complicated to organise and deliver, but the Centre has a very extensive archive of materials, produced by a beautiful study at the Roslin Institute in Edinburgh, which is proving invaluable in assessing test performance on preclinical samples, which are the ones you most want to test. The preclinical samples seem to have less of the marker of infection than those from clinical affected animals and the same is probably true in humans. This presents another hurdle because samples from preclinical human cases are almost non-existent although there are a few from blood donors who went on to develop the disease.

The net result of this has been slow progress and a number of honourable failures, but as of now there seem to be two blood tests, one in France, the other in America which look very promising. One of them is very near the stage of CE marking, which is part of the process by which a diagnostic test is approved for use in Europe.

Tests for CJD are even more difficult than most because the infected state is almost entirely without any marker of infection before symptoms develop

# The PRION 2016 Conference

**Each year, there is an international conference on prion diseases. PRION 2016 was held in Tokyo, Japan, over three days in May**

Professor Richard Knight

**Around 350 or so delegates attended PRION 2016, including clinical doctors, pathologists, laboratory scientists, veterinary experts along with people working in public health and others.**

Included in these others are representatives of patient/family support organisations, led by the CJDISA (the CJD International Support Alliance), of which the UK CJD Support Network is a member. The PRION meetings traditionally give some time in the meeting for the CJDISA to present to the scientific and medical audience, allowing laboratory researchers an insight into the devastating personal effects of prion disease and, thereby, giving them renewed motivation in their work.

There is insufficient space here to discuss the content of this conference

in detail and, inevitably, some of it – such as discussions of structural protein chemistry – is technically challenging to the non-expert (and, as a clinical neurologist, I include myself here!) However, topics other than prion protein chemistry included:

- Animal prion diseases (including the emerging Chronic Wasting Disease of deer species)
- Surveillance of human diseases in different countries
- Pathogenesis (how neurons become damaged in prion diseases ie the actual mechanism of disease)
- Advances in clinical diagnosis
- Possibilities for treatment
- The expansion of the prion concept.

I will pick out three of these for a little further discussion.

Firstly, there has been a lot of progress in diagnostic methods. As there are

separate article in this newsletter on the topic, I will not go into further detail. However, the underlying theme concerns so-called ‘amplification techniques’ that allow detection of very low levels of abnormal prion protein and how they may be efficiently and reliably used in clinical practice to obtain earlier and more accurate diagnosis.

Secondly, the expansion of the prion concept refers to the fact that the pathological processes that characterise prion disease may be relevant to other diseases such as Alzheimer’s disease or Parkinson’s disease and there were a few interesting presentations on this area. The precise relationship between these rather different clinical illnesses remains uncertain but potentially interesting.

Treatment is, of course, the most important area for those affected directly or indirectly by these diseases and it was notable that this conference had a specific section on treatment.



One thoughtful talk discussed the reasons why treatments that have looked promising on the basis of laboratory work have not, so far, led to meaningful benefit for patients, and how we might make better progress in the future. Underlying many of the presentations and discussions, the questions concerned the specific disease aspects that treatments should target and whether successful therapy might require targeting more than one disease aspect. For example, we know that normal prion protein is necessary for disease, and is converted into an abnormal form, that abnormal prion protein accumulates in disease tissue, that proteins (normal or abnormal) can be cleared from tissues and that neurons (brain cells) become sick and die by various possible mechanisms – but which of these is the best treatment target or targets?

There is natural concern that effective treatment is most likely with early diagnosis and, even better, with pre-symptomatic diagnosis. This is particularly relevant in genetic prion disease where an individual carrying a relevant mutation may be well for years before developing clinical disease. One interesting presentation concerned an Italian treatment trial in which members of a genetic prion disease family are being treated with a drug (Doxycycline) to see if it delays or prevents disease – a trial that will need to run over a number of years before any definite result is available (projected possibly to be around 2023).

Progress in science and medicine is often slow but it does take place and you can, I hope, take some comfort from the fact that so many researchers meet annually to discuss their continuing work on these diseases that have affected your lives so terribly.

*PRION 2017 is in Edinburgh in May 2017 and the CJDISA and your own Support Network will be represented there, to present your concerns and interests and to learn of the progress being made.*

# The CJD Foundation Family Conference Washington 2016

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Mrs Blaire Smith-Bathgate, Senior Nurse & National Care Co-Ordinator,  
National CJD Research & Surveillance Unit

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## **On the 8th of July this year I joined with families of those affected with Creutzfeldt-Jakob Disease (CJD), scientists and health care professionals to attend the yearly CJD Foundations Family Conference in Washington DC.**

The conference runs over a weekend and consists of workshops, bereavement support, lectures and presentations. Whilst there is well-structured formal programme, the weekend also allowed for families to spend time speaking with other families and professionals about their own experiences of dealing with CJD. Many of the conversations lead to ongoing support from both the professionals and the foundation.

There were several lectures given by scientists sponsored by grants from Family Memorial Grants and the Foundation itself. There was a strong focus on the role of animal models in identifying and testing treatments, using these models to explain how to delay or prevent the onset of prion disease by reducing the production of the prion protein and how to achieve that in practice. One of the issues raised was in relation to treatment and the poor translation of trials from animals to humans and the effect of the blood brain barrier i.e. whether drugs will actually cross the barrier to have any effect on the brain. The role of the Cerebral Spinal Fluid (CSF) test, RT-QuIC (second generation) is considered highly specific and sensitive for CJD and a great aid to an earlier

diagnosis. Whilst these presentations were very scientific, every effort was made by the presenters to explain their work to a wide and varied audience.

The families who attended had relatives who had suffered from genetic, sporadic and iatrogenic forms of the disease. Some members of the genetic group were aware of their status and other not, some were awaiting results. There were interesting discussions of the pros and cons of having this information. Some of these families were regular attenders at the conference. The people I met were very supportive of each other and said they relished the opportunity to speak freely with others who had been down a similar path. Some others attenders were very recently bereaved. Some had taken some time to feel able to attend but for all it was a time of re-visiting the past but equally looking forward to a more positive future.

A presentation from the Centres for Disease Control and Prevention (CDC) questioned the rate of the disease that are commonly mentioned within the US i.e. 1–1.5 per million of the population per year, although rates of up to 2 cases per million are not unusual. A more compelling figure they quote is that there is 1 CJD death for approximately every 6,000 death overall in the US each year (with others quoting that figure as 1 in every 5,000 deaths).

The final (and fourth) day of the conference on Monday is spent by

the remaining family members visiting Capitol Hill to attend advocacy visits. An advocacy training session is held on the previous day by a legislative aide to help prepare families who attend. The primary purpose is to meet with members of Congress and their staff and educate them about CJD. The key request is to ask for Congressional support to continue the National Prion Disease Pathology Surveillance Centre (NPDPSC) funding, thus allowing them to deal with increasing autopsy costs and continue to develop more efficient detection levels of prion diseases.

The Foundation has spent much time and effort trying to evolve with the speed of changes in technology. They have recently started a teleconference support group. A guest speaker gave an overview of a preselected subject and answered questions put to them by the programme director. The phone lines are opened up for families to ask questions they may have. This process

has been very successful in providing yet another route to provide support for families at whatever stage they are at in the CJD path and is easily accessible for all. The Foundation also uses Family Workshops which visit various states and allow for those unable to attend the annual conference with support and information. Given the size of the country this has worked very well. A Facebook group is also utilised. Whilst they receive a grant from the Centres for Disease Control and Prevention, there are numerous funding activities throughout the US organised by families in the form of golf outings, running marathons, dinner parties, bake sales and numerous other activities to support their work.

From a personal point of view there are two main things I took away with me from Washington this year. Although I work and have worked within the CJD field solely since 1998, it is events like

this when scientists and professionals from around the world get together, make presentations and the following discussions and debates, illustrates the many people out there working on the numerous aspects of this disease. The work on diagnostic testing to treatments it really is quite incredible.

The second thing which struck me was that although we complain about our health system (NHS) and its faults are widely documented, at least we have something in place. Some of the people I spoke to told terrible stories of being unable to get medical help, waiting for tests, poor communication issues and a lack of state support when it came to care. Issues with undertakers and no funding for autopsies also added to their problems. Many felt isolated and alone. Without a diagnosis they had nowhere to go for support such as that provided by the Foundation.

## UK CJD figures

**Recent numbers of deaths of definite and probable cases in the UK.  
Figures from the National CJD Research and Surveillance Unit in Edinburgh**

Year	Sporadic	Iatrogenic	Genetic	vCJD	Total
2010	84	3	7	3	97
2011	91	4	14	5	114
2012	93	5	11	0	109
2013	106	2	8	1	117
2014	98	3	11	0	112
2015	104	0	4	0	108
2016*	69	1	4	1	75

\* As at 3 October 2016

Source: <http://www.cjd.ed.ac.uk/documents/figs.pdf>

## Fundraising for CJSN – a piece of cake!



Brenda Blake organised a cake sale day in memory of her brother John Saunders, who died in 2015 of sCJD.

Brenda wrote, 'Lisa Saunders, who sold the cakes on the day, has her own cake making business in Swindon. She brought her cakes and cupcakes, as did several other people who also made cakes for the event. People could have a cup of tea or coffee plus a choice of a cake to go with it.

'A great day was had. People were asking if they could buy whole cakes or a selection of cupcakes, which was good (they obviously liked them!)

'We all worked well as a team. Stephanie and Lorna took orders for the drinks in the kitchen with Ivor and Serena washing up. People gave us raffle prizes, so Bridget ran a raffle, with Linda and Margaret helping with ticket folding. Angela, Sheila and Thirza helped by selling handmade cards. Meanwhile, the rest of us, David, Sue, Jackie, John and I, were making sure that everyone had what they wanted.

'John would have been proud to see us all working so hard – but what fun we had doing it, and we may do it all again next year. When we added all the money up, what a surprise we had – £400!'

## Yard sale

To mark the first anniversary of her husband John's death on 3 September Sue Saunders held a yard sale. Tables were set up under the car port and luckily it only rained for the last 20 minutes. Sue was very grateful to the many neighbours and friends who

attended the sale and helped her to raise £130 for the CJD Support Network in memory of John. She was particularly grateful to Daryl and Marcia who came and helped to set up at 8.30 in the morning and worked tirelessly until the end.

## Donations in memory

Heartfelt thanks to the families and friends of those below for donations received in their memory between August 2015 and 1 September 2016

Jasmine Ancona  
 Anna Bellerby  
 Barbara Betty Bennett  
 Peter John Closey  
 Chris Collins  
 Pat Conlon  
 Peter Cotton  
 John Cutting  
 Daniel (Danny) Patrick Doherty  
 Jacqueline Downes  
 Antony Escritt  
 Geoffrey John Garvey  
 Carole Gibbins  
 Maxine Hayes  
 Margaret Hipwell

Robert Hirschorn  
 Tony King  
 Danny Martell  
 Peter Milton  
 Maureen Jesse Lillian Mitchell  
 Clive Morgan  
 Margaret Mary O'Sullivan  
 Mark Pope  
 John Alfred Saunders  
 Stuart Taylor  
 Trevor Paul Wallis  
 Alan Walsh  
 Patrick Wellings  
 Barbara Wright  
 Linda Young

### My brother Mark

Karen Goodall

After several months of GP appointments, various tests and hospital visits my brother, Mark, was finally diagnosed with vCJD in August 2015 at the age of 36. We first noticed that there was something wrong around a year prior to this. Mark had become quite withdrawn and had problems with his balance, which meant he became confined to a wheelchair, the kind of thing you would want for a six foot two 16 stone scaffolder! I found that his short term memory was poor but bygone events such as nights out and meeting old acquaintances were recapped as if they occurred the previous day. As Mark's condition declined we were unable to have conversations with him and on Tuesday 23rd February 2016 Mark passed away surrounded by his family. During the illness Mark was not aware of his condition and the ultimate outcome which was a blessing.



## Management Committee 2016



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



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



**Karen Goodall**  
Karen's brother died of vCJD



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



**Professor Simon Mead**  
Simon is a neurologist working at the National Prion Clinic.



**Andy Tomaso**  
Andy's mother Carmelina died of Genetic CJD in 2007



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



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



**Gillian Turner**  
**CJD Support Network**  
co-ordinator



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



**Anita Judd**  
Anita's family is affected by Genetic CJD. Anita and her cousin Stuart run the Inherited CJD Support Group.



**Roger Tomkins**  
Roger's daughter Clare died of vCJD.



**Kate Dahill**  
Kate's aunty died of Sporadic CJD

We are planning a series of telephone conferences in 2017 providing information and support. Please keep a look out for further information on our website [www.cjdsupport.net](http://www.cjdsupport.net)

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 a tablet or smart phone. These useful pens cost £2 plus p&p each and you can buy them by contacting the Network at the address below.



**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 complete the form below and post to the CJD Support Network, PO Box 346, Market Drayton, Shropshire TF9 4WN. Membership is free, but we welcome donations.

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