CJD SUPPORT NETWORK

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Newsletter

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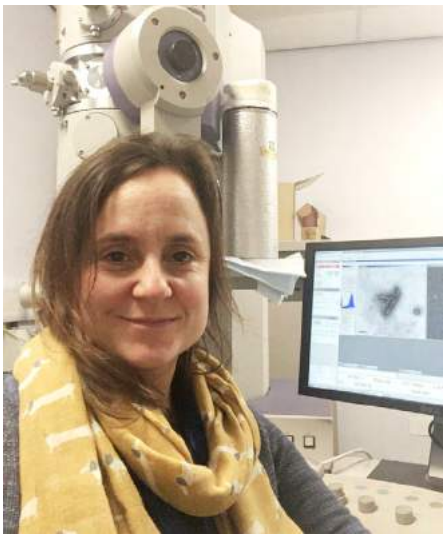
CREUTZFELDT-JAKOB DISEASE SUPPORT NETWORK NEWSLETTER

Investigating the structure of prions

A report on the presentation at the CJD Support Network Family Support Meeting 2017

Dr Cassandra Terry, UCL Institute of Prion Diseases, MRC Prion Unit



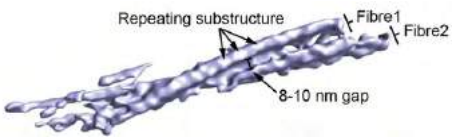
What is prion disease?

In prion disease one of the body's own proteins called the prion protein (PrP) somehow misfolds into a misshapen, rogue form that clumps together and accumulates in the brain. These misfolded rogue proteins (or prions) spread throughout the brain (and sometimes other parts of the body) by converting more and more normal prion protein into the rogue form resulting in progressive damage to the nervous system. The prion protein can misfold into several different shapes or 'strains' resulting in different forms of prion disease with different characteristic features. Our research is focused on revealing the structure of these rogue proteins to help us better understand the disease, develop better diagnostic methods and hopefully new treatments.

Isolating prions from diseased brain

For many years scientists studied artificial non-infectious versions of misfolded prion protein made in the laboratory as there was no easy way to isolate genuine rogue proteins from brain to see what they looked like. However, after much effort supported by funding from the Medical Research Council (MRC) we managed to develop a way of isolating prions from diseased brain so that we could study their structure directly. We confirmed that the structures we isolated were authentic by labelling them with a PrP–specific antibody (to show they were made of the prion protein) and measuring their biological activity using specialised cultured cells.

What do prions look like?



I had to use a really powerful microscope (that uses electrons instead of light to illuminate the sample) to look at the rogue proteins we isolated. This microscope can magnify up to 100,000 times, meaning really small particles such as proteins can be viewed directly. I collected a series of tilted 2D images of the rogue proteins by tilting the platform inside the microscope. I then used a computer program to build up a 3D structure from all the different 2D images I collected. This was the first time that prions had been viewed in 3D. My images revealed that prions are made of twisted pairs of short double helical prion protein fibres, a structure that is common to all the prion strains I examined. I then compared them to artificial PrP fibres we made in the laboratory (that don't cause disease) to see if they differ. Disease-causing prions from brain have a far more complex structure than the artificial fibres, which are composed of only a single double helical fibre. There is clearly something very important about the structure of prions that make them capable of causing disease.

Moving forward

Following further support from the MRC and UCL we will soon have our own dedicated microscope that will allow us much more time to image prions in detail. We are now using more complex methods that don't require the sample to be stained (stain improves contrast in the images it can also cover fine features). We are particularly interested to see what is down the middle of the prions to understand what is keeping the two paired fibres together. This may explain why they are so robust and difficult for the body to destroy. We also want to compare different prion strains to see precisely how their structures differ. Understanding the structure of prion strains is important not only for prion diseases but also for understanding other neurodegenerative diseases such as Alzheimer's disease, which involve the spreading and accumulation of other misfolded proteins in the brain. ■

CJD Support Network
Chairman's report 2017–2018

Professor Richard Knight

Welcome to the latest edition of our newsletter. Over the last year, the Support Network has continued its general support for patients and families affected by CJD but has also undertaken some new developments.

Research funding

The first of these has been the provision of a fund to support two research projects. This is in the process of being advertised and we hope to attract applications which will then be reviewed by a panel of experts. The successful applicants will be invited to talk about their work at the annual Family Days. In this way, members of the Network will be able to see how their fund-raising helps to contribute to better understanding of CJD and researchers will have direct contact with those affected by the disease on which they work.

Family Support Meeting

The second development concerns the Family Day itself. Aside from being able to include talks from researchers we are directly supporting, we have decided to trial a change in the meeting structure for our November 2018 day. The meeting will begin on Friday evening with a Reception and a Dinner with the opportunity for participants to stay overnight in the hotel or locally. The main meeting will be on Saturday as usual, but the Friday event will allow more time for families, the committee members and experts to get to know each other and exchange experiences. If this is successful, we will make it a regular event.

Personal data and confidentiality

The Network's Management Committee is reviewing its confidentiality policies and practices. As we hope you will appreciate, we have acted in a very careful manner in relation the information we receive about individuals and their circumstances. We have no intrinsic concerns about our practice, but as you are no doubt aware from the news, there have been several concerns about privacy and confidentiality in certain areas. In the wake of these concerns, there are legislation changes and we need to be absolutely certain that our organisation fully meets all the formal, legal, requirements.

We plan to discuss any further implications of this at the November 2018 family meeting.

The whole committee and I hope you feel the Network is continuing to serve you and we would welcome any suggestions as to how we might do this better. Please come to the November Family Support Meeting if you are able to do so; we look forward to seeing you there.

Our helpline 0800 085 3527

We continue to receive a steady number of calls to our 24 hour helpline.

Calls include those relating to newly diagnosed cases of CJD, those where people have been told they are at a heightened risk and the worried well (who have often become worried after Googling CJD or their symptoms!) Some people just want to talk to someone who will listen.

Where we are not able to provide an answer to the question being asked, we are usually able to refer the caller to a person or organisation who will have the answer.

We get a number of calls from people wanting to donate to the Network or fundraise for us. In this latter case, we can offer advice and provide T-shirts and collecting boxes if that is helpful.

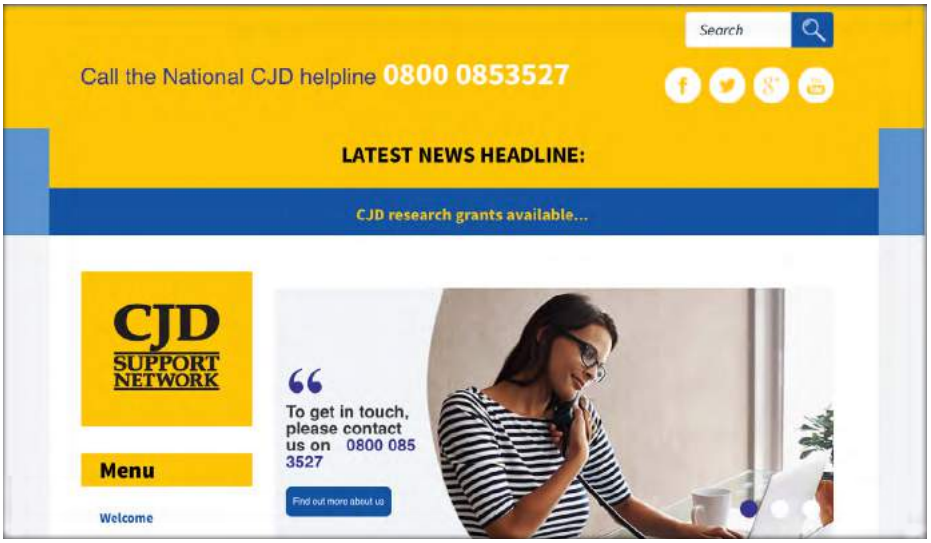
We are now getting more email enquiries via our website 'Contact us' page, which is proving to be a useful means of contact.

Anyone wishing to speak to us should telephone Freephone 0800 085 3527, or email support@cjdsupport.net

Updating leaflets, brochures and guidelines

We are currently reviewing and updating several of our leaflets and booklets, to ensure the information in them is up to date. These new versions will replace older versions and be made available on our website or from the CJD office.

What do you think of our website?



We constantly review and update our website at **www.cjdsupport.net** to keep it up to date and relevant as a resource for patients, families, carers, fundraisers, professionals, students, researchers etc.

If you find the website useful, we would be delighted if you would tell us. Equally if you cannot find things you would find helpful, or you find that there are errors on the site, please let us know and we will do our best to correct things.

Shortly after publication a copy of this newsletter will be added to all the previous newsletters on the website. These can be viewed or downloaded, as can all our leaflets and booklets.

Representing the Network views at NICE

The National Institute for Health and Care Excellence (NICE) is currently developing updated guidelines and protocols on 'Patient safety and reduction of risk of transmission of CJD via interventional procedures'. The CJD Support Network has a representative on this committee, to ensure that patients, families and carers are at the centre of their decisions when they are made.

The current NICE guidelines, which were published in 2006, can be seen at www.nice.org.uk/guidance/ipg196

General Data Protection Regulations (GDPR)

This new EU-wide law comes into force on 25 May 2018 and applies to all organisations and businesses who hold or process personal data of a European citizen. This therefore applies to the CJD Support Network.

You will almost certainly have had communications from several organisations you have dealing with, such as banks and insurance companies, regarding this.

This new law does not itself change any of the existing Data Protection Regulations, but places a responsibility on all organisations to be able to show that they are complying with these regulations.

Our privacy policy

Our Privacy Policy statement, which sets out how we comply with the requirements of the new law, is available to view on our website. It will be pointed out to anyone who is giving us any personal details, for example over the phone.

We feel however, that it would be helpful to identify some key points from our policy.

Key points in our privacy policy about personal data

- The CJDSN holds the minimum personal contact information about those we have dealings with, to enable us to provide the best possible service to everyone who contacts us,
- It will only be held with the person's permission
- This information will never be shared with third parties (without explicit permission, or where required to by law)
- Anyone can ask what information we hold relating to them and ask for it to be corrected or deleted completely. In the last situation, we would of course be unable to contact them in the future, for any reason

Great benefits from attending a Family Support Meeting

Sandra and Mike Walshe

In August 2007 my sister in law Kay died of Sporadic CJD, leaving us all devastated and unable to make any sense out of what had happened to her over the last five months.

Three months later my husband and I attended the family support meeting, not something Mike would have even contemplated under normal circumstances. Initially his thoughts were: why would I want to sit down in a meeting all day with people I don't know, what will I be expected to do and do I really want to talk to strangers about loosing my sister while I'm still feeling traumatized and upset.

But these were not normal circumstances. No one he had spoken to knew anything about CJD, the doctors treating Kay had not met anyone with this disease before, it felt like a closed shop when it came to finding out any information other than through the CJD Support Network helpline – which really did feel like a lifeline at the time.

On reflection now Mike thinks it was the best thing he ever did to start the healing process of coming to terms with his loss. He had felt that for months nobody he spoke to knew anything about CJD and then he found himself sitting in a room full of people who all knew something about the disease. It felt almost serene.

Everyone who attends the meeting is affected by CJD in some way or other, families with someone newly diagnosed, people who are living with the disease or the threat of it and sadly those who have lost someone either recently, or some time ago. Everyone has a common bond and without exception benefits from the mutual support of one another. People generally arrive feeling a little anxious and uncertain as to what they are letting themselves in for, but always leave with a smile, better informed and reassured that they are not alone.

Experts in the field of CJD give talks about things of interest. They are on hand to answer questions either in a group, or discreetly over coffee or lunch. Some people just want to listen and take it all in, but here is never a better opportunity to raise the un-answered questions and talk to people in the same position, in a warm, friendly environment.

We are looking forward to the extension of the family support meeting over two days this year. It will be good to have more time to spend with families who we have got to know over the years.

Birmingham

On a lighter note for those coming who wish to take in some sightseeing, I feel duty bound as a Brummie to tell you a little of what Birmingham has to offer.

You're never short of retail therapy in The Bullring, Grand Central and The Mailbox, or you could enjoy a wander round Brindley Place's stylish canal side bar's and restaurants. Birmingham is home of the world-famous Symphony Hall and the iconic, historic Town Hall, or you could take in a show in Theatreland and visit Chinatown. The Shakespeare Memorial Room and Secret Garden for views over the city are situated in the Library of Birmingham, or maybe you could catch a tram to the Jewellery Quarter. These are all on the doorstep of the Burlington hotel, formally known as The Midland Hotel, that is referenced in the popular TV drama *Peaky Blinders*.

CJD Support Network
Family Support Meeting 2018
16–17 November in Birmingham

At the request of families who attend our Family Support Meetings who ask us to allow more time for families to mix and talk with other families who have had a similar experience of CJD, we have arranged a two-day meeting.

The Family Support Meeting will be over the weekend of Friday 16 and Saturday 17 November 2018. The venue will be the Burlington Hotel, New Street, Birmingham.

The meeting will start with a welcome reception on Friday, followed by dinner. On the Saturday the meeting will follow the pattern of previous years, with a variety of expert talks and family discussion groups. Tea and coffee at registration and a buffet hot lunch will be available.

There is no charge, the Network will fund the Friday reception and dinner and the Saturday meeting including food and drink. If you wish to stay overnight at the Burlington Hotel we have arranged a special accommodation price which you will need the code for to book at the reduced price. There are also a choice of different hotels close by. A member who attended last year told us that he stays at the Central Travel Lodge in China Town which is very close to the Burlington and where you can get a very good deal, especially if you book early. He also says the car parking at the hotel is very much cheaper than most.

I do hope you will decide to join us. As you will see from some of the articles in this and earlier newsletters, families find attending both helpful and interesting and it gives the opportunity to meet other families who have had a similar experience of CJD to you.

If you wish to attend you will need to register by telephoning Gillian Turner on 01630 673993 or the helpline on 0800 0853527.

Lovely people at the Family Meeting

My name is Annette. I lost my husband Mick to sCJD on 6 May 2017. He was aged 72 years. I was at a loss after he passed, feeling angry and alone thinking I was the only one. Blaire from Edinburgh University's Research and Surveillance Unit kept in touch – this offered some comfort that he wasn't just another number.

Then I saw on the CJD Support Network UK site that there was to be a family support meeting in Birmingham in November 2017 so I contacted them by telephone and spoke to a very helpful lady named Gillian, I told Gillian I would attend the meeting however I was very nervous and unsure what to expect from going, I had no need to be though as everyone was very welcoming

Annette's husband Mick

just being able to speak to others about CJD as they know what you have gone through and are still going through and knowing that you are not alone.

I met some lovely people. If I hadn't attended the Birmingham Family Support Meeting that would not have happened. We had a lovely relaxed meal on the Friday evening where we could all chat and get to know each other a little this put my mind at ease before the meetings on the Saturday. I felt so comfortable to be able to talk about Mick and his life.

I will be attending the Birmingham family support meeting again this year with our daughters, hopefully they will get as much out of this as I did. I am looking forward to meeting up again with friends old and new. I would advise anyone that has any misgivings and may feel apprehensive about attending a family support meeting to come along meet others and not feel isolated or alone.

My long, long journey with CJD

Graham Blades

I attended the 2017 family support meeting after a 20-year absence. During those 20 years and more, I had devoted my time to caring for my beautiful wife Janet and our young family. Janet was originally diagnosed with sporadic CJD at the young age of 27.

As a founder member of the CJD Support Network I had kept in touch with Gillian Turner and the team up in Edinburgh. Sadly, early in 2017, Janet passed away after a long struggle with her illness.

LEFT **Graham's wife, Janet, who died in 2017 after he had cared for her for 29 years**
RIGHT **Graham and children**

I had always said that perhaps sometime in the future I would become involved again. The Family Support Meeting last November far exceeded all of my expectations. From the warm friendly welcome at registration

at 9.30am until close at 3.30pm, the day had flown by.

This event has become very well honed by Gill and the team on the Management Committee. The 'ice breaker' concept early in the proceedings helped enormously – putting all family members at ease by demonstrating there were others in very similar circumstance to yourself. Be it the type of Prion disease (sCJD/vCJD/Genetic/GSS or Iatrogenic etc) and also what part of the UK you are from.

I met someone in very similar circumstances to myself who only lives a few miles away. I have spoken with him several times since.

There were many excellent presentations given by experts in different fields – from talks on prion disease, current research projects, advice on social care, bereavement and grieving, all of these experts were more than happy to answer any questions from family members.

Above all of this though the best part is meeting people who have gone through, or are going through the same experiences as yourself. Whichever form of prion disease has affected your family we all have more in common than we do that is different and therefore can share our knowledge and experience in a way to hep and comfort each other knowing we truly understand each other in a way others cannot.

I understand from Gillian Turner that the 2018 family support meeting will be over two days. I would encourage any family member to attend this excellent event, but I fully understand if you are hesitant in attending as it can be difficult to take that first step.

I am sure that if you attend you will take away something positive, from something that has had such a negative impact on your life.

Bereavement: The Grieving Man

A summary by Anita Tipping of the talk by **Ian Leech** at the 2017 Family Support Meeting

Ian Leech and his wife lost his daughter through cancer. He is a community engagement and supportive care manager at St Giles Hospice. He related the difficult times he and his family went through, his coping mechanisms, his relationships, mood swings, and happy memories. His grieving started at the point of diagnosis.

Mr Leech's daughter was diagnosed with Hodgkin's Lymphoma and passed away nine months later in 2008. Father and daughter were close. Both loved going to football matches and both laughed at the same things. Like any parent losing a child he was at his lowest.

Grieving is unique with everybody, individuals grieve in their own way. There is no right way or wrong way to grieve.

Mr Leech felt suicidal for months following their loss and his relationship with his wife came close to separation. He felt he needed to be strong at his weakest. He sought help and medication helped him to feel normal again. Busying himself by writing a book helped him deal with his grief. Understanding and recognizing their differences saved his and his wife's relationship and marriage.

Although he hated cycling, he went on a cycling training course because he was planning to cycle from London to Paris and back, as a way of giving back to the community who helped him and his family during their loss. As a result of this grueling cycle ride, he found out he enjoyed cycling and that he needed cycling as his therapy.

He attended his daughter's best friend's graduation day and did a charity cycle ride to thirty different football grounds, in memory of his daughter's 30th birthday.

During his talk, Ian Leach identified the following key points when grieving,

- Manage yourself and your health.
- Never be afraid to acknowledge why people do not know what to say.
- Talk and listen to those closest to you.
- Do not put a time limit on your grief.
- Do not let anyone tell you how to grieve.
- Do not be afraid to cry or show emotion.
- Learn to be kind to yourself and get on with your hobbies.

New Director of the National CJD

Support Network committee member Beth Marsh caught up with **Dr Alison Green**, Reader in Biochemistry and recently appointed Director of the National CJD Research & Surveillance Unit (NCJDRSU) in Edinburgh

Hi Alison, could you start by filling us in on what your role as Director of the unit involves?

I've got to have an overview of the direction of research being done by the NCJDRSU, ensuring our research is asking the relevant clinical and scientific questions and that we are gathering as much data as we can to answer those questions. We have a senior management team which has an epidemiologist, a clinician, a neuropathologist and myself and we work together to make sure the unit works as a whole. We also manage the finances, represent the unit on university committees and liaise with the Department of Health (DOH) and the Advisory Committee on Dangerous Pathogens (ACDP).

It sounds like a complex role!

The unique thing about the NCJDRSU is that it has a wide range of staff that include neurologists, neuropathologists, scientists and epidemiologists that all work together to provide clinical, scientific and public health information relevant for CJD surveillance. As Director I have to make sure that all areas of investigation work together in a coherent fashion.

I'm surprised to hear that the research and surveillance unit has so much of a clinical focus…

For surveillance we still need a clinical input and we need to see the patients and their families. We need to make sure we're not missing cases so that we've got a good idea of the range of presentations. This means that we can give advice when people ring up and say 'I've got a patient with this kind of movement disorder' and we can say 'we did have a case that presented like that', or 'we've never seen this before'.

What role does the unit play in supporting families and patients?

We have two research nurses whose aim is to ensure that families get the support and care that they need from local organisations. The nurses get to know families and have a wide experience of the range of things people can feel and be frustrated about. It's such a rare disease that a lot of the clinicians who see patients initially may have never seen a patient

Research & Surveillance Unit

with these symptoms before. I think they find it helpful when they get clinical advice from our neurologists, so we work to support the families and also local clinicians. It is frightening for everybody because people become so ill so quickly.

CSF (cerebrospinal fluid) research is an area in which you've had many achievements. What do you think has been your most influential research finding so far?

I think that the RT-QuIC test has been the most influential. The test was developed by Dr. Byron Caughey in Montana and one of his colleagues, Professor Atarashi. When they suggested that it could be used to detect abnormal prion protein in hamster CSF, I thought 'well if we can do it in hamsters, why can't we do it in human patients?' Adapting and developing a test that's now part of the diagnostic criteria has been the biggest thing for me.

So if a patient has a lumbar puncture now, that's what happens to the CSF – the RT-QuIC?

Yes. We also still do the 14-3-3 which is the traditional test. There are some cases where the RT-QuIC may be negative and the 14-3-3 positive and vice versa. We're interested in those cases and would like to find out why. We've also adapted the RT-QuIC test to look at another abnormal protein, alpha-synuclein, that's found in patients with Lewy body dementia and Parkinson's disease.

How did you become interested in prion/neurodegenerative disease research?

I did biochemistry and pharmacology, then an MSc in Clinical Biochemistry. I worked in a clinical biochemistry hospital for about ten years and was always interested in proteins as markers of disease. Neurologist Dr John Patten was sending CSF samples off to the National Hospital for Neurology to test for Multiple Sclerosis (MS). He said 'would you be able to set that test up in your lab?' So that's where I started getting interested in CSF proteins. And then (in about 1994) I got a job at the National Hospital to do a PhD looking at CSF proteins in MS. During that time the BSE epidemic was evident and in 1996 they announced the first cases of vCJD. There was a test called 14-3-3 emerging and again somebody said 'why don't you do it?', so we set it up. Then Bob Will (Director of the Research & Surveillance unit at the time) came to give a talk. I said to him 'you know we can do 14-3-3 here?', and he said 'well that's great because we're sending the samples off to the States at the moment, so why don't we send them to you?' – that's how I got involved in CJD.

What are some challenges of working in prion research?

The most challenging thing is to find out what is the component is that causes the neurological damage and find out how we can stop that from doing what it's doing. With prion disease it takes twenty years for anything to happen and that's another challenge. You need to have records that go back years. This is where the DOH have been very supportive, they have funded us for a long time and we've managed to collect data going along and this is a great resource. A lot of researchers are looking at the way we've been collecting data and thinking 'this is what we should do for other diseases'.

What's the best part of your job?

The best thing about my job is when somebody finds something new, and says 'oh look, come and see this!' and you think 'wow – that's great!', that's the exciting bit.

What are the new and current developments in prion research?

We have Chronic Wasting Disease (CWD) emerging in Europe and recently prion disease has been identified in camels. That's fascinating in itself – how it all emerged, how it expanded, how CWD arrived in Europe, where camels suddenly developed this disease. There's also a lot of work looking at sensitive methods for detecting abnormal prion protein, we've got abilities to look at replication of the proteins. So there are new analytical tools and we've got this emerging animal disease, those are the new things that are making us look at prion disease in a new way.

Who have been your biggest inspirations and why?

Bob has been pretty inspirational and Richard (Knight) has got a great way of looking at complex problems. The other person would be Jane Goodall. She did animal research by watching chimpanzees in the wild, it was observational research and it was a novel approach. In a way that's what we do in surveillance. In

addition to our basic research we also observe patients with CJD. You gain an awful lot of understanding about diseases by observing and talking to people and families, putting the data together and looking at it over years. And, personally, my grandmother because she was such a fantastic woman. My grandfather was killed in the second world war and she brought my mother and uncle up by herself. She worked relentlessly to make sure they had everything they needed. Then they both got married and there she was at 50 thinking 'what do I do now?'. She decided to get a job in New Zealand and make a new life for herself. She travelled all round the world and as a child I'd get postcards from these fantastic places and it was just 'WOW'. She was a big personal inspiration and from her I learnt that you can get on and do anything, you just have to really go for it.

What do you think are the benefits of researchers and scientists getting out of the lab and engaging with families affected by CJD?

From the point of view of scientists and researchers it's about not losing track and not getting so wrapped up in the fact that you're looking at this protein or doing this test. When you see and understand the impact of the disease on the families it brings home what you are trying to do. We are trying to help understand this disease and to be able to give people an answer to questions they ask us.

I think from the families' point of view it stops researchers being seen as sort of mad people that don't have contact with the real world. The research we do is complicated, it's difficult, and a lot of the time people think you've been studying this for ten years and you've got no more answers. Engaging with families means we can say 'yes, but these are the reasons why and this is what we've learnt along the way.'

I think the way forward with research into these diseases is to have a coherent group of researchers. We're all in it together. So you have the contribution of the patients, scientists and clinicians and everybody sees it moving forward bit by bit. The families of CJD allow us in at a very difficult time. We appreciate the time that people have given us to sit down and answer questions when really they may not want to. We really appreciate that. The NCJDRSU has a CSF biobank and we ask family members if they would mind if the residual CSF sample from their family member is stored for research and nearly everybody agrees. It's fantastic because when we did the work on the RT-QuIC we had such a big store of CSF samples that we could analyse immediately. So we published a paper in 2012 and got it into the diagnostic criteria in 2017, but we only set it up here in 2010. We could only do that with the family support with giving consent for samples to be stored. That's the benefit of us working together.

The NCJDRSU is holding a family day on 6th September 2018 in Edinburgh and all family members are welcome. For more details, please contact Elaine Lord at: Elaine.Lord@ed.ac.uk

Social work

Peter Simcock and Jill Manthorpe

The features of CJD are such that both health and social care professionals often need to be involved in supporting the person and their family. Social workers are typically part of the team or network supporting people with CJD, offering assessment, access to social care and support services, care co-ordination and direct therapeutic work. However, the rarity of CJD means that it is often outside the experience of most social workers.

In the 1990s, a CJD Support Network survey identified a distinct lack of awareness, knowledge and understanding of the condition amongst the social work profession. In particular, there appeared to be limited awareness of the need for urgent responses to requests for social work support, in view of the rapid disease progression common in CJD. These findings led, in 1998, to the production of the first set of good practice guidelines for social services professionals working with people with CJD, by Derek A Biggs, then Operations Manager at Cambridgeshire Social Services and a pioneer in practice development in this area. The guidelines were subsequently amended and re-published in 2003.

The legal and policy framework for adult social care in England has changed significantly since the publication of the amended version of the guidelines. This is predominantly as a result of

and CJD: updating the guidelines

legislation, such as the Mental Capacity Act 2005 and the Care Act 2014 and also increased personalisation, a policy agenda across public services which centres on offering people more choice and control over their care and support. It is therefore timely that the guidelines are revisited, and we were delighted to be joined by Prof Simon Mead (Consultant Neurologist and Clinical Lead) and Selam Tefsamichael (Clinical Nurse Specialist) both from the UK National Prion Clinic, in a unique collaborative project to refresh and update them, drawing on practice wisdom, research intelligence and CJD expertise.

We explored the literature on social work and social care practice in the field, identifying themes to inform the structure of the updated guidelines. These themes were placed in the context of the new legal and policy frameworks, and the experiences of the staff at the UK National Prion Clinic and a first draft was developed. We are especially grateful to colleagues from the National CJD Research and Surveillance Unit in Edinburgh and from the CJD Support Network, who offered constructive comments, feedback and additions to the earlier drafts. Peter was keen to talk about the work on the new guidelines at the CJD Support Network 'Family Support Meeting' in November 2017 and received very helpful suggestions for additions, from those attending.

A near final version was completed in December 2017 and the Department of Health and Social Care will publish the final version later this year. The new guidelines provide social workers with advice and information about a range of topics, including the different types of CJD, the role of the National CJD Care Team and National Prion Clinic specialist nurses, assessment and care planning for people with CJD, and matters relating to the funding of care. The new guidelines also provide advice on supporting carers and family members, recognising that a diagnosis of CJD may impact on the whole family. The information in the guidelines is supported by a series of illustrative, anonymised case studies, drawn from the work of the National Prion Clinic and contact details for other sources of support and information are included at the end.

The updated guidelines have been supported by the Chief Social Worker for England, Lyn Romeo, who acknowledges their potential to equip social workers with the knowledge and skills they need when working with someone, likely for the first time, with CJD. However, it is our hope that the new guidelines will prove to be a useful resource, not only for practitioners, but also for individuals and families affected by CJD.

Peter Simcock is Senior Lecturer in Social Work at Birmingham City University and a PhD student at King's College London. Prior to working in social work education, Peter worked for two local authorities as a social worker, senior practitioner and team manager, predominantly in adult disability services.

Jill Manthorpe is Professor of Social Work at King's College London, Director of the Social Care Workforce Research Unit and NIHR Senior Investigator Emeritus. She works on a number of studies funded by the Department for Health and Social Care, provides assistance on workforce matters to local councils, NHS bodies and user, carer and patient groups, and provides expert advice to the care sector and to other NHS Arm's Length Bodies about care services and workforce initiatives.

BSE, Variant CJD and the Appendix III Study

Professor Richard Knight
National CJD Research & Surveillance Unit, Edinburgh

It has been generally considered that the period of risk of contamination of human food in the UK by material from from bovines affected by BSE (Bovine Spongiform Encephalopathy) ran from 1980 to 1996. Prior to 1980, it is thought that BSE infective material did not enter human food production. After 1989, following measures to protect human food, the risk was felt to be small and, indeed, to date vCJD (variant CJD) has not been identified in the UK in anyone born after 1989. In 1996, further protection measures were put in place and, therefore, this later date has been taken as the end of significant BSE/vCJD UK risk to humans via diet.

Despite estimations of quite significant amounts of potentially infective material entering human food between 1980 and 1996, there have, thankfully been relatively few cases of vCJD in the UK. This is, of course, not to ignore the tragedy of those who were affected – it is just that the magnitude of the disaster could have been much greater. This apparent mismatch between the potential for infection and the actual number of people infected has a few possible explanations:

1. That there is a fortunate protective barrier making bovine to human transmission unlikely and this is considered to be an important explanation.

2. That the potentially long incubation period of prion diseases means that further cases of vCJD will arise. This is likely to be true but expectations are that relatively few people will be affected.

3. That humans can become infected, but never actually become ill – remaining in a subclinical infected state

The last two possibilities are of concern as they imply that there are individuals in the general population who are silent 'carriers' of infection who might infect others by, for example, by blood donation and contamination of surgical instruments. Indeed, transmission of vCJD via blood donated by individuals in the pre-clinical stage of vCJD has occurred, although in only a very few instances.

This potential for human-to-human transmission has, understandably, been a significant public health concern in the UK, with various protective measures put into place.

An obvious question coming out of these concerns is: how many people are sub-clinically infected? Attempts have been made to answer this question and have relied on the idea that BSE infection in humans begins with silent infection in tissues like the tonsil and appendix well before any neurological symptoms appear. The basic method has been to examine tonsils or appendices, removed in ordinary surgical operations, for the abnormal disease-related prion protein that is deposited in prion disease.

There have been two major studies undertaken that examined mostly appendix material and have been, informally referred to as the Appendix I and II studies. These studies found abnormal prion protein in a number of samples suggesting that around 1 in 2,000-4,000 of the UK population were silently infected (and, indeed, most would never be expected to become ill). It is important to note that these studies were anonymised so that it is simply not known from whom the appendix specimens came.

To date, variant CJD has not been identified in the UK in anyone born after 1989, when measures were introduced to protect human food

There was, however, a further question, namely: does the presence of abnormal prion protein in the appendix definitely result from exposure to BSE? This is a very reasonable assumption but one that is not beyond doubt. A further study was designed – Appendix III – looking at two further population groups: appendices removed from individuals during surgery performed before 1980 and appendices removed from individuals born after 1996. These two groups being unexposed to dietary BSE risk, as discussed above. The expectation was, therefore, that no positive results would be found in either group. However, positive results were found in both and the numerical analysis found no statistically significant difference in the rate of abnormal appendices in the three time groups: pre-1980, 1980-1996 and post-1996.

There are two broad explanations for this unexpected finding:

1. That abnormal prion protein in the appendix is not always an indication of BSE infection. However, if this is so, it is not clear as to what it would indicate.

2. That the dietary exposure period in the UK was more extensive than supposed – beginning before 1980 and extending after 1996. However, from what is known about the BSE epidemic, it is not straightforwardly obvious as to how this could be.

The full details of the Appendix III study are not yet published. Discussions are ongoing as to how to interpret the results as they stand and further research is planned to see if the questions raised can be answered. However, at the moment, uncertainty surrounds some of the assumptions that have been made about BSE and vCJD in the UK.

Deaths from definite or probable CJD

Year	Sporadic	Iatrogenic	Genetic	vCJD	Total
1990	28	5	0	-	33
1991	31	1	4	-	36
1992	45	2	6	-	53
1993	36	4	7	-	47
1994	53	1	9	-	63
1995	35	4	5	3	47
1996	40	4	6	10	60
1997	59	6	6	10	81
1998	64	3	5	18	90
1999	62	6	2	15	85
2000	48	1	3	28	80
2001	58	4	6	20	88
2002	73	0	5	17	95
2003	79	5	6	18	108
2004	50	2	6	9	67
2005	67	4	13	5	89
2006	68	1	9	5	83
2007	63	2	11	5	81
2008	84	5	6	2	97
2009	78	2	8	3	91
2010	85	3	6	3	97
2011	91	4	14	5	114
2012	94	5	11	0	110
2013	106	2	8	1	117
2014	99	3	11	0	113
2015	105	0	4	0	109
2016	118	1	6	1	126
2017	108	0	8	0	116
2018*	35	0	3	0	38
Total	**1962**	**80**	**194**	**178**	**2414**

*As at 4 May 2018 Source: NCJDRSU website www.cjd.ed.ac.uk

Tough Mudder challenge

Holly and Sacha Evans and a group of friends (right) successfully completed the Tough Mudder event last September and raised nearly £5,000 for the CJD Support network.

Welcome support

CJD Support Network Treasurer Andy Tomaso (below left) received a cheque for £3,352.18 from Gary McAllister at Brampton Heath Golf Club. Gary nominated the Network as the charity for his Captain's Golf Day, in memory of Rita Ann Brown.

Donations in memory

Heartfelt thanks to the families and friends of those below for donations received in their memory by the CJD Support Network between April 2017 and May 2018

Gordon Barwick
Anna Bellerby
Barbara Betty Bennett
John Blaylock
Denise Anne Cleeve
Chris Collins
Pat Conlon
Paul Croft
John Cutting
Stephen Roger Diamond
Peter Evans
Steven William Goddard
Chris Gaukroger
Pauline Harding
Beryl James
Wynford Jones
Michael Gerard Kiely
Philip and Denley Osborne
Rhoda Lillian Paulley
Susan Pow
Geoffrey Victor Sanders
David Harry Skelton
Sylvia Storer
Mrs Surrey
Mark Webb
Margaret Wellings
Patrick Wellings

Research funding call 2018

The UK CJD Support Network (CJDSN) invites applications for research grants for the year 2018.

Applications may be for:

- Total funding of a new small research project.
- Partial funding of an existing or proposed research project.

In either case, the maximum sum will be £25K.

The research project can be in any area concerning CJD and related prion diseases, including laboratory, clinical and care aspects, but preference will be given to projects concerning human disease, especially diagnosis and treatment.

An application form can be found on the CJDSN website. It should be completed and submitted by 30 June 2018

Applications will be reviewed by an expert panel, including members independent of the CJDSN. Any reviewer who has a conflict of interest with respect to any application will be excluded from that particular review. The reviewers will report to the CJDSN Management Committee who will make the final decisions.

Applicants will be informed of the outcome by 31 August 2018.

Successful applicants will be required to provide progress reports to CJDSN and also to attend the CJDSN Family Day (usually in Birmingham, in November each year) to present their work to a family audience. Any resulting presentations and publications should, of course, acknowledge the CJDSN financial support.

Further details and instructions for submission can be obtained from the CJDSN website **www.cjdsupport.net** by going to News and Events / Latest News.

Management Committee 2017–2018

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

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

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

Karen Goodall Karen's brother died of vcjd.

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

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

Kate Dahill Kate's aunty died of Sporadic CJD

Beth Marsh Beth works in psychopharmacology. She lost her father to sporadic CJD in 2016 and joined to support young people.

Gillian Turner CJD Support Network co-ordinator

Blair Smith-Bathgate Blair is a National Care Co-ordinator and Senior Nurse at the National CJD Research & Surveillance Unit in Edinburgh.

CJD Support Network free helpline number 0800 085352

Feel free to call us whether you want to find out some information or just want to talk to someone.

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

☐ Please tick to agree to us keeping this information on file. We need this to contact you and will never give it to a third party except with your explicit consent.

CJD SUPPORT NETWORK
Contact: Gillian Turner, CJD Support Network, PO Box 346, Market Drayton TF9 4WN
Telephone 0800 0853527 · Email gturner@cjdsupport.net · Website www.cjdsupport.net
Charity number 1097173