CJD SUPPORT NETWORK

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Newsletter

Issue 24
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CREUTZFELDT-JAKOB DISEASE SUPPORT NETWORK NEWSLETTER

The CJD Support Network: 20 years of providing support

- Networking families & professionals
- 13,000+ helpline calls answered
- 20 family support events
- Factsheets, newsletters, podcast
- Worldwide visitors to website
- Research sponsorship
- Eight large conferences
- Representation to Government
- Support for CJD networks abroad
- Guidance on CJD to WHO

In December 2015, the CJD Support Network celebrates 20 years of providing practical and emotional support for families and professionals affected by CJD

Beginnings

The CJD Support Network formally started in 1995 as part of the Alzheimer's Society, which had attracted three years of Department of Health funding to develop a support service for patients and families affected by all strains of CJD.

Gillian Turner was appointed as National CJD Co-ordinator. Her role was to develop a carers' group for people who contacted Alzheimers about CJD. At this time CJD was a rare dementia which was very little known and little or no information about the disease was available in written form which could be easily understood by lay people.

Our first steps were to establish a 24-hour helpline and to commission factsheets and other pieces of information on all the different strains of CJD.

Variant CJD

It was only a few months later when the CJD world changed from a rare, little reported disease, to a high profile disease, through the identification and diagnosis of what was first called new variant CJD. This became variant CJD (vCJD).

In March 1997 the CJD Support Network held the first conference on CJD at Warwick University. We were worried that we would not attract enough delegates to make the event successful. We needn't have worried, not only was it a sell-out, but we attracted 30 television and radio crews and a large contingent of reporters from all over the world.

Being part of the Alzheimer's Society, with Clive Evers as our CEO, had many advantages at the beginning, as we were able to call on all their specialist staff. On the day of the Warwick conference we were so thankful for the Alzheimer's Society press department, headed by Simon Denegrie, who handled all the press crews and managed all their demands for interviews.

CONTINUED PAGE 2

The CJD Support Network: 20 years of providing support continued

Independence

We continued to grow, but in 2003, due to lack of funding for the CJD Support Network project, the Alzheimer's Society said they could not continue running the Network. Without any funding, a small team led by Dr Angus Kennedy, our then Chairman, Gillian Turner, Clive Evers from the Alzheimer Society and a few family committee members including Anita Tipping (Secretary) and Mike Curtis (Treasurer), fought for independent charity status and in 2003 we were officially registered as an independent charity, run from an office in Gillian's home. We applied for, and were fortunate to secure, a year's start-up funding from the Department of Health.

In the same year Gillian was awarded the MBE in the Queen's New Year Honours List for her work with the CJD Support Network.

After our Department of Health funding ended, we went through many worrying times, even sometimes doubting whether we could maintain all our services. However, through sound financial management and wonderful financial support from donations and by fundraisers, we are now on a strong financial footing. However with no statutory funding, we will continue to need this financial support from the public.

Our role

The CJD Support Network's main role is to provide practical and emotional support for families and professionals affected by CJD. Our main service is our 24-hour helpline, where we have answered over 13,000 helpline calls. However we have undertaken other complementary tasks over our twenty years.

Other networks

Over the years, we have given support and advice at the start-up of similar support networks around the world including Japan, USA and Germany and we are a member of the International CJD Alliance.

Committee membership

In the UK, the Network has been present, as an expert lay representative, on several Government committees. This has enabled us to ensure that the patient and carer's views are considered in administrative, political and medical decision making.

Other activities

Since 1995 we have hosted eight large conferences, held a family support meeting each year and produced 23 newsletters. We have produced CJD guidelines for Social Workers and Nursing, as well as prompting the Department of Health to produce CJD Health Guidelines. In 1999 we were asked to help the World Health Organization (WHO) to produce world guidance for CJD. We have also sponsored a piece of research on sporadic CJD at the National Prion Clinic.

Website

Our website gives everyone access to our specialist information and advice on all strains of CJD. This includes downloadable factsheets on the different strains of CJD, a CJD podcast and CJD audio information, as well as our newsletters. There is also a fundraising section. Our website is an integral part of our support to families and professionals, attracting five to six thousand unique visitors from all over the world each month. With this in mind, to mark our 20th year, we have commissioned a new website, which should be up and running by the end of the year.

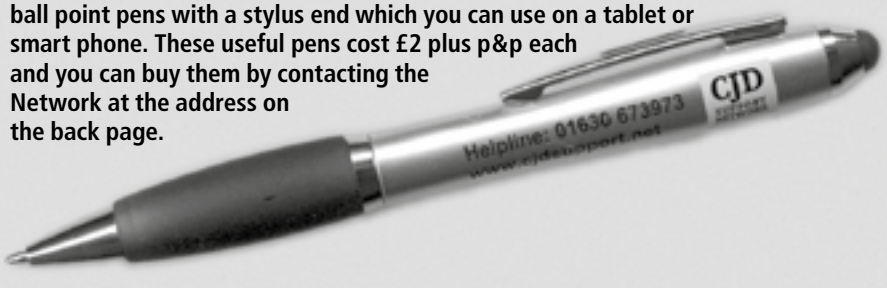
Support and the future

Along the way we have met and worked with some wonderful families, carers, committee members, and professionals, too many to list here. We would like to thank them all for helping the CJD Support Network to get to where we are today, twenty years after our start, and to thank them in anticipation for their continuing support in the years ahead.

This year, sadly, we have had to say goodbye to Derrick Biggs. Derrick was appointed by the Association of Social Work Directors in 1997 to act as the CJD link person. Derrick wrote the social work guidelines for CJD and has worked tirelessly since his appointment to ensure social workers were informed about CJD and family needs and has helped many social workers to access the help that families required if a referral was proving difficult. We thank Derrick for all his help over the past 17 years and wish him a happy retirement, and look forward to working with his replacement. See the back page for a statement from Derrick.

Pen with stylus for tablets and phones

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 on the back page.



Geraldine Hillier entered the 2015 London Marathon and raised £5,046.53 in memory of Aynsley Lambert

Brian Southall and his son entered the Sunderland 10K and raised £387 in memory of Dorothy Mutch

In memory of my aunt

Steve Marshall

My dear aunt, Teresa Mary Cox Millis, was so tragically taken by Sporadic CJD on the 17 November 2013. Teresa was 57 years old and full of life.

I decided to to take part in the London to Southend Bike Ride in her memory to raise money for the CJD Support Network. I raised £1,750. Since I took part in the bike ride I have become a lot fitter and lost weight due to carrying on this great sport.

I would advise anyone who is thinking of raising money for this great charity to do what I did and start cycling.

Lastly I would like to say a huge thank you to all the people who sponsored me for the event.

Steve Marshall at the finishing line in Southend

Can you help us this year to raise money?

In 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 back page).

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.**

My grandfather

Jenna Sendall

Jenna's grandfather Mick Rumsby is on the right

The start of this year shocked my family and me, as my grandfather (69) was taken into hospital. Scientists rushed over from Edinburgh to the Norfolk and Norwich NHS to do some tests, which resulted in his accurate diagnosis.

At first, the hospital staff were sure that he had suffered from a stroke. It was three months until they gave him his full diagnosis of CJD. 'One in a million' the doctors said.

Everyone that has known my grandfather would have told you about his bravery, strength and incredible love for his family and friends. For him to deteriorate the way he did and as quickly as he did was unnatural and heartbreaking to see. Worst of all, there wasn't a single thing we could do to stop it.

Michael Glenn Rumsby, 'Mick', sadly passed away on March 26th 2015 in a care home close to home. He was surrounded by loved ones and was peaceful.

I wanted to find out more about CJD and I wanted to raise awareness, therefore I contacted the CJD Support Network and arranged a sponsored walk event.

On Sunday 31st March, I was accompanied by my older sister Jordan and a number of others to walk 10 miles. I had arranged a local route, including the village where my grandfather had lived.

We started off in North Walsham Town Centre and headed out through Swafield, then to Trunch, through Mundesley and Knapton, then back into North Walsham.

The five hours seemed daunting but everyone strived and not a single walker gave up! It was set to be a rainy and windy day with thunder showers throughout. Luckily, we only had a single shower and mild winds.

Together we walked, laughing and made awareness in our yellow attire. We even got sponsors on route!

I am thankful for everyone who took part and sponsored, also the kind help and advice from the CJD Support Network team.

Together we have made a difference.

Donations in memory

Heartfelt thanks to the families and friends of those below for donations received in their memory between May 2014 and June 2015.

Alan Bedford
Alfred Green
Anita Elizabeth Hughes
Ann Bellerby
Aynsley Lambert
Barbara Wright
Barrie Albert Papworth
Barry Walton
Brian Morrisey
Christine Hills
Christopher Little
Clive Morgan
David Leslie Webber
David William Youngerman
Diane James
Diane Jones
Dorothy Mutch
Fay Dickeson
Geoffrey John Garvey
Gordon Barwick
John Cutting
Lynda Harry
Margaret Anne Utting
Margaret Ann Lee
Micheal Glenn Rumsby
Nigel Robinson
Pat Conlon
Paul Marshall
Richard G A James
Rosemary Grace Harrison
Stephanie Mary Austrin
Steve (Rob) Robinson
Stuart Taylor
Susan Hardacer
Teresa Mary Cox Millis
Trevor Hughes
William Walter Wright
Wynford Jones

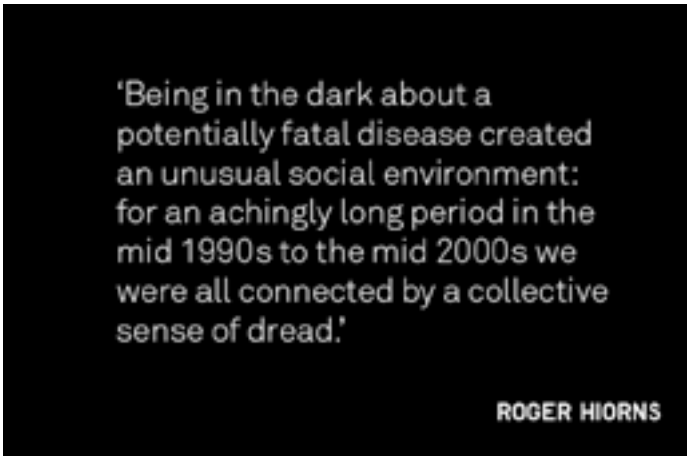
For any fundraising news of your own – or if you would like to name someone on our memorial role – or to make a donation, please contact Gillian Turner at the CJD Support Network. Contact details are on the back page.

History is Now: 7 Artists take on Britain

Exhibition at Hayward Gallery, London
10 February – 26 April 2015

History is Now was an exploration of 70 years of British cultural and social history. Seven UK based artists of different generations and backgrounds – John Akomfrah, Simon Fujiwara, Roger Hiorns, Hannah Starkey, Richard Wentworth, Jane and Louise Wilson – were each invited to curate distinct sections of the exhibition, looking at particular periods of cultural history from 1945 to the present day.

As part of the exhibition, Turner Prize nominee (2009) Roger Hiorns, created a presentation exploring the biological and social impact of bovine spongiform encephalopathy (BSE) and its human equivalent, variant Creutzfeldt Jakob Disease (vCJD). Working with leading scientific experts in the field, including Professor John Collinge of the MRC Prion Unit, University College London, Hiorns has undertaken an investigation into the histories of BSE, vCJD and other prion related diseases, together with related developments in biomedicine, agriculture, husbandry, food production and consumption. Supported by a People Award from the Wellcome Trust, Hiorns' presentation took the form of a multi-layered display containing works of art, scientific objects and research,



government reports, press coverage and research materials. This was the first time that this subject matter had been explored in an exhibition format.

We understand that vCJD remains a current issue within public health and that being sensitive toward those who have been affected is as important as raising public awareness.

CJD Support Network invites you to the **Annual Family Support Meeting** 21 November 2015

The CJD Support Network management committee are pleased to announce our 2015 family support meeting will be on Saturday 21 November at the Burlington Hotel, New Street, Birmingham.

Further information will be distributed nearer the date, but I do hope you will put Saturday 21 November in your diary. The Family Support Meeting is one of the highlights of our year, as it is lovely to meet new families in person and catch up with old friends.

CJD SUPPORT NETWORK

Every year we receive lovely letters from families, saying how helpful and enjoyable these meetings are. It is a great chance to meet and talk with other families who have had a similar experience of the disease and to put those unanswered questions to CJD experts.

This year the program will include talks from Professor Richard Knight and Professor Bob Will from the National CJD Research & Surveillance Unit. Joining us also on this special occasion will be Clive Evers, who was the CJD Support Network's first Chairman and link with the Alzheimer's Society. Dr Simon Mead of the National Prion Clinic will also be at the meeting.

The Prion 2015 International Conference

Professor Richard Knight

Each year, there is a major international conference concerning prion diseases; the most recent – Prion2015 – was held in May at Fort Collins, Colorado. Around 500 scientists and clinicians (from around the world) attend these meetings, where research and observations are shared and discussed.

Those affected directly by these diseases are represented by the International CJD Alliance, formed from the support organisations of different countries including the UK's CJD Support Network. The attending scientists hear direct accounts of the personal effects of prion disease and say this helps to motivate them in their further work towards understanding and, eventually, curing these illnesses.

It would be difficult to provide a detailed summary of the four days of presentations and impossible within the space of this article; instead, I will give an account of the main themes with some emphasis on one or two particular aspects.

There was discussion of prion diseases of animals, as well as those of humans. Quite aside from matters of animal health and agricultural policy, such diseases are important from a human viewpoint. No one needs to be reminded of the human consequences of BSE in cattle and studying animal prion disease can give insights into human illness. Indeed, some of the real progress in prion disease has resulted from the fact that animals suffer naturally from such illnesses. At this conference, the animal disease emphasis was on CWD (Chronic Wasting Disease) which affects various cervid species (deer and elk). This emphasis on CWD partly reflects the fact that it arose in, and principally affects, North America. However, it is of general concern as it has spread rapidly across many USA states and appears to be a rather contagious disease in deer populations. There is theoretical concern that it may be transmissible to humans but there is no current evidence that it has done so.

It might surprise readers that one fairly big theme concerned yeasts. However, yeasts, like all living things, contain proteins and some of these proteins show changes that parallel the changes seen in animal and human prion protein. Because yeasts are grown and studied easily in the laboratory, they have been used as a convenient model for prion disease study, without any of the ethical concerns relevant to animal experimentation. There are interesting results from yeast experiments that aid thinking in the prion disease area but it is not always clear how relevant these are to human disease in the final analysis. Incidentally, for those of us who like beer, bread and other baking, the existence of such prion-like behaviour in yeast proteins poses absolutely no risk to human health (of course, excessive beer and baking consumption may do so, for entirely different reasons!).

Another major theme was the relationship between diseases such as CJD and other neurological diseases of ageing (Alzheimer's, Parkinson's, Motor Neurone Disease etc). All of these illnesses can occur sporadically or genetically, involve misfolding of specific proteins and are characterised by degeneration of brain neurones (nerve cells). There appear to be important common disease mechanisms in these otherwise rather different diseases. The increasing overlap of these fields of research is likely to benefit understanding of all these illnesses. Prion diseases have one important distinction in that they are actually or potentially transmissible (from animal-to-animal, animal-to-human and human-to-human), whereas this has never been demonstrated for the other diseases. The exact reason for this difference and the precise nature of the infective agent in prion disease remains uncertain.

I would select two broad themes from the human prion disease presentations.

1: The precise mechanism of prion disease

We know that the normal prion protein misfolds into an abnormal form that is deposited in brain tissue and we know that brain neurones become sick and die; the latter being the cause of symptoms. What is still not clear is the relationship between these two events, or, to put it more starkly, we don't fully understand why the neurones stop working properly and degenerate (and this, after all, is the core disease question). There were a number of presentations of work trying to address this matter and, bit by bit, a clearer picture does seem to be emerging. Without going into detail, and with the risk of over-simplification, I will select four elements that might give at least a sense of research in this area.

Firstly, on the basis of the known fact that normal prion protein (designated PrPC) misfolds in prion disease and forms an abnormal prion protein (designated PrPSc), original considerations tended to centre on whether depletion of PrPC (via loss of its normal function) or accumulation of PrPSc (via a toxic effect on neurones) was the fundamental disease mechanism. This consideration has tended to give way to the notion that there are intermediate protein forms between the normal PrPC and the easily identified tissue deposits of PrPSc and that these intermediates may be the critically neurotoxic elements.

Secondly, there has been some move away from the notion that a form of abnormal prion protein is simply neurotoxic in itself and evidence that it may be an interaction between this and PrPC ie that the normal form of prion protein is involved in mediating a toxic effect of abnormally folded protein. It has long been known that PrPC is an essential requirement for developing prion disease.

Thirdly, proteins are dynamic things that are constantly being made, folded, unfolded and destroyed; mistakes can be made in this process and there are cell mechanisms that deal with these mistakes. Research has focussed on these 'quality control' mechanisms in order to understand why misfolded prion protein might cause problems.

Finally, there has been increasing interest in how the immune system (the defence mechanisms against infection) might be involved in disease production and this has been considered in other diseases as well (such as Alzheimer's).

2: Better and quicker diagnosis

One problem facing patients, families and clinicians is that of getting a secure and speedy diagnosis; CJD can present in ways that are similar to other diseases and many of the diagnostic tests have been non-specific (ie not based on unique prion protein abnormality that is the hallmark of CJD etc). There have been quite a few developments in recent times including a technique called RT-QuIC. This test is able to detect reliably very small amounts of abnormal prion protein and has proved extremely useful in the diagnosis of sporadic CJD (the commonest human prion disease). Further development of this technique was discussed at the Prion2015 conference that may lead to a more sensitive test (being positive in more cases) and one that is much quicker to perform. Other developments may lead to applications somewhat wider than that of sporadic CJD and, indeed, considerations have been given as to how this sort of technique might aid diagnosis in other diseases such as Alzheimer's.

Treatment

As will be obvious, I have not mentioned treatment. There are quite a few potential therapeutic approaches that have arisen from research, including antibody treatments (directed against normal or abnormal forms of protein). Understanding the mechanisms of neuronal dysfunction and death better should help in the development of treatment. I do not think there was anything very new presented at the Conference, but you should be assured that a great deal of energy and time is being put into work towards possible treatments.

Living with a rare hereditary condition

Anita Thompson

My family are affected by inherited prion disease (genetic CJD); we discovered this devastating condition in the family in 2000. I was given a copy of the following piece by a special person who counselled me through very difficult times.

The piece was written by a mother of children at risk of Huntington's disease, another rare hereditary condition. Living with a rare hereditary condition like genetic CJD or HD in your family is devastating and can be very overwhelming. It's always there, always in the background, affecting the things you do and the decisions you make in life.

Discovering genetic CJD in my family has been life changing for me and other members of my family. I sometimes think of the time when we were unaware of it.... However, I think myself and other members of my family have changed our lives for the better, because of it; with the realisation of how precious life is.

EDITOR *We welcome Anita as our newest member of the CJD Support Network management committee.*

The Stranger

By a mother from New Zealand Huntington's Disease Scene who has children at risk of Huntington's disease

I never knew *HD*. He just burst into my life one day unannounced. As the door slammed I knew he was here to stay. 'You've got no business here' I told him. But he just smirked and sat down in the big cosy chair by the fire.

As the flames roared in a burst of yellow and orange, I watched out of the corner of my eye to see what he was going to do next. He just sat there, unknown, unannounced, as though he had been here in my house all along, as though he was part of our family.

'Look, HD maybe you've got the wrong house, There's a really nice couple that lives down the street, got your eyes, I could show you the way if you'd like'.

'No thanks, I'm staying for good, we go way back. Besides it's nice to catch up with kinfolk'.

So HD settled in and we had no say about this unwelcome guest.

The other night I went dancing with *Anger*. She wore a red dress which twisted and swirled as she did the two-step in her stiletto heels. She was really easy to dance with, but when it came time to leave, she didn't want to go.

The music played on till 2am and I was tired so I left *Anger* there and went home alone, only to find HD waiting up for me. I try to ignore him sometimes, pretend he isn't here in my house, playing with the kids and sharing himself around. But his attitude remains one of indifference, it's like he doesn't care what we think, he just carries on regardless.

I was busy cleaning up after lunch on Tuesday when there was a knock on the door and it was my old friend's *Joy* and *Compassion*. They felt I'd been out of touch lately and came round to remind me we had a date on Friday for coffee and a chat.

I was so pleased to see them and we laughed about the time we'd played soccer with the Autumn leaves at Anzac Park, which was funny really, because it was a time of friends coming together to remember. When it was time for them to go, I gave them a big hug and said I'd see them soon. As I showed them out I could see HD's shadow lurking behind the kitchen door.

It was like that sometimes, you couldn't see him but you knew he was there. His tall, dark features were hard to miss and the foxy grin of his told tales of sneaking around. He seemed to bring out the worst in me somehow. I'd try to be polite and make conversation but I never invited him in so I would always put on a mask when he was around,

it was the only way I could cope for a while.

Yesterday the sun was shining, it was the first day of spring and the first daffodils were out in my back garden amongst the weeds. *Complacency* had taken over the gardening duties and she wasn't doing a very good job, I'm thinking of replacing her with *Nurture* whom I've heard is a whiz at such things. It would be so nice to see the tall poppies making a show at Christmas.

This evening *Longing* and I sat on the porch and watched the last remains of the day sink slowly below the horizon. It was still warm outside and it was kinda nice sitting there not saying much while the moon crept in to light up the night sky. *Longing* never likes to talk much, he just sits there staring into the distance with a wistful look on his face. I asked him if I should ring up *Comfort* and ask her around so that we could listen to music together but he wasn't at all pleased with the idea and said he was happy just the way he was.

It took me a long time, but after talking to *Father Hope* after mass last Sunday, I decided to forgive HD for coming into my life so unexpectedly and shattering our peaceful existence.

I threw a party to celebrate and invited all of my old friends; *Courage*, *Love*, *Inspiration* and *Harmony*. *Beauty* even turned up for a while wearing her pink satin dress and matching shoes. There was lots of laughter and fun as we popped the champagne and waltzed around the room.

Some uninvited guests turned up for a while, I found them gossiping in the room with HD. I suspect he invited them in, but would never admit it. He introduced them as *Suffering* and *Grief*. They wore black jeans and army-style shirts, spilling their beer and swearing a lot so I called *Security* over and asked him to kick them out.

It was well after midnight when everybody left. *Pleasure* was the last to go. I was sorry to see her leave but she said she'd be back again soon.

HD is part of the family now, he introduced me to *Change* and *Resignation* after the final match last Tuesday. The score was nil all. Although I didn't invite him in, we've got used to having HD around and we don't argue as much as we used to.

Sometimes I hear him on the phone ringing up *Fear* so I make an excuse and go out. Most times though we get along okay though. I've got to know him pretty well now, he's no longer a stranger so that's nice, isn't it.

Next summer he's decided to come on holiday with us. I'm not sure about that, so I've asked *Fun* to come along just in case.

Inherited Prion Disease Support Network

The Inherited Prion Disease (IPD) Support Network was founded by two cousins to develop peer support for others affected by IPD.

We hope that our community is a place to help to signpost, share information, join in discussions and most of all feel supported and not alone.

Please come and visit our website www.inheritedpriondiseasesupport.org

Planning or taking pot luck?

Facing the future with a genetic disease

Alastair Kent, Director, Genetic Alliance UK

'Prediction is difficult, especially when it is about the future' the Danish physicist Neils Bohr once notably remarked. But for people living with the threat of a genetic disease hanging over them the opportunity to plan can be central to their ability to cope with the present and to face the future with a degree of confidence.

Of course, we can never be totally confident that we have dotted all the *I*s, and crossed all the *T*s, but given the necessity to face a future with a genetic condition looming over you, most people will want to make some form of plan or coping strategy.

The first step to planning is an accurate diagnosis. Without this then you struggle to escape from the working of blind chance. If you do not know what is wrong then you cannot know what to expect. This is incredibly stressful for everybody – the affected or at risk person, his or her family and the professionals charged with the provision of care and support. Everything changes when a diagnosis is made.

Information is key to this. Without accurate, comprehensive and comprehensible information that you can rely on then any plans are likely to have foundations built on sand. The internet has been a huge boon to many in creating routes to know, but not everything on the internet is trustworthy, some of it is complex and technical, and much of it may be written in language that is difficult for a lay person to take on board – 'it's English, but not as we know it'.

Navigating the maze is much easier when you have a guide. Support groups such as the CJD Support Network can help sort out the wheat from the chaff in what can often seem like a blizzard of conflicting views and opinions. When trying to understand a rare condition, peer to peer support can be a real lifeline. With common, better understood, conditions help is available from local sources – the GP, the district nurse etc. Families with rare conditions must look further afield, as they will often be the only ones in an area with that particular condition. Where the condition results in the person affected looking 'odd', or behaving differently then they may feel isolated or even stigmatised. Contact with others in the same situation can help reduce this feeling of isolation and provide helpful explanations which will help disarm the suspicions of friends and neighbours, enabling local awareness and possibly creating the opportunity for a local support network.

Support groups for ultra-rare conditions (and the CJD Support Network is an excellent example of this) have often found what I call the 'Amazon' approach to developing supportive networks helpful. When you live with the threat of a rare condition it can be difficult to know what to expect. In this situation there are, to quote Donald Rumsfeld, 'things we know we know, things we know we don't know, and the unknown unknowns'. Like Amazon, support groups can offer suggestions – 'people in your position have found the following things helpful', or 'have found the following questions useful in getting information from their doctor/genetic counsellor etc'. This approach has the benefit of being helpful without being prescriptive, because people can always say 'thanks, but no thanks' if the suggestion doesn't work for them in their specific circumstances. It is also easily developable and modifiable in the light of experience and new knowledge.

So, to complete the circle, we can see that Neils Bohr was only partially correct. We can take steps to improve the predictability of the future by working together and pooling our experiences, helping build the critical mass that will open us up to planning, and reduce the risk of pot luck being in the driving seat!

Reducing the risk of iatrogenic CJD from medical devices: a personal view

Professor Andrew Smith, NHS GG&C and University of Glasgow, Scotland

A summary of the presentation Andrew Smith provided to the CJD Support Network annual meeting in Birmingham 2014 on his experiences from a Scottish perspective of measures taken to reduce the risk of transmission of CJD via contaminated medical devices

Introduction

It is vital at the outset to note that there have been six cases of CJD transmission from neurosurgical devices (four cases from neurosurgical instruments and two from stereotactic needles). This is in the face of several million lifesaving and changing surgical interventions. However, there is also compelling laboratory data to support recommendations for vigilance and measures to improve the cleaning and sterilization of instruments used in all forms of surgery. The laboratory data suggest that the infective agent of vCJD is difficult to wash off from instruments and any residual infectivity is less likely than ordinary bacteria and viruses to be killed by steam sterilization, the most common method for sterilizing surgical instruments.

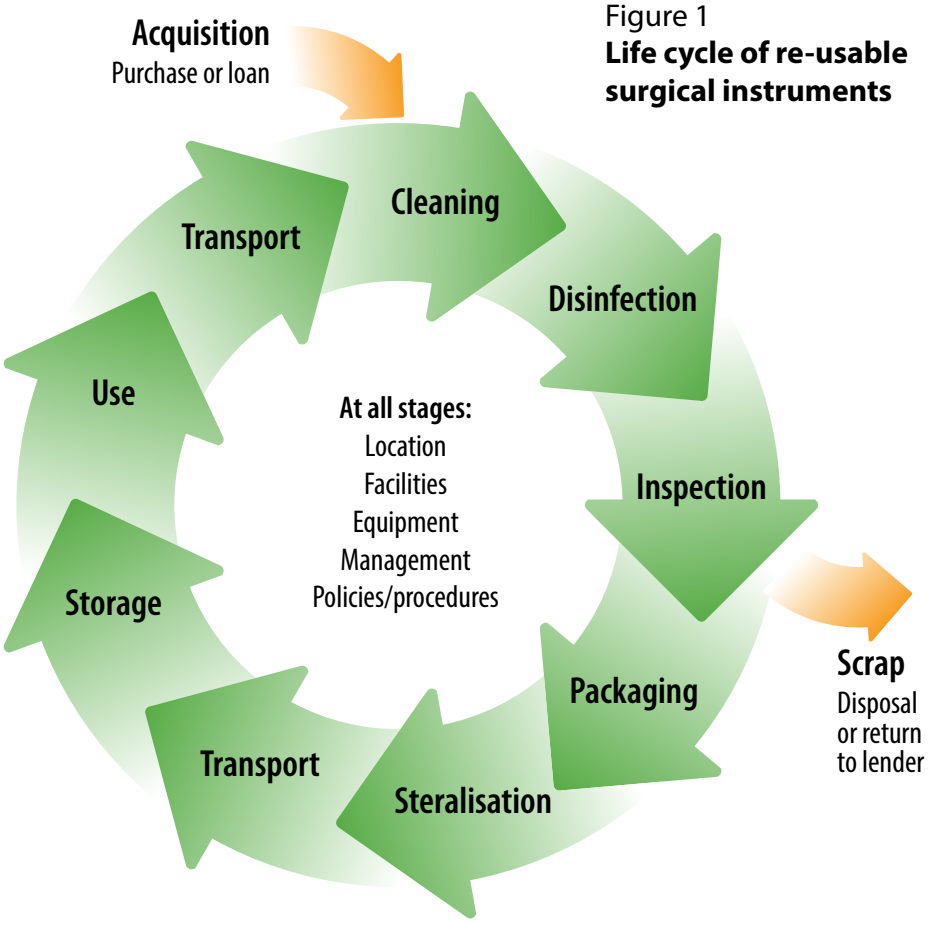


Figure 1
Life cycle of re-usable surgical instruments

The surgical instrument decontamination cycle

The term instrument decontamination is now used in place of disinfection or sterilization of surgical instruments. This is because it is now widely accepted that there are several stages important to ensure that medical devices are fit for use. Figure 1 summarises the key stages in the life cycle of re-usable surgical instruments. Each stage is typically regarded as a critical control point for entry to the subsequent stage. Or put more simply if you screw up one stage, the whole cycle falls apart! Some instruments are not reprocessed but are used once only and then discarded. Obviously this represents zero risk from the cross-infection perspective but carries significant burdens in terms of waste, environmental impact, transportation and storage logistics. Although there are some interesting efforts to recycle the material used in surgical instruments into other (non-medical) uses and I would expect to see a greater use of investment in this approach in the near future.

Historical perspective

One of the first reported investigations into the performance of NHS hospitals in the decontamination of surgical instruments was published in 1958

and consisted of an investigation into 'sterilizing practice' in six hospitals in England. It is more affectionately termed the 'yellow peril', the yellow refers to the colour of the cover and peril from the nature of its findings! This report found several significant short comings in the safety of surgical instrument reprocessing and one of its recommendations was that it was 'essential to replace the current 'laissez faire' approach by a structure in which there is a defined responsibility, proper equipment and training'. This study was followed up by a Medical Research Council Working Party who published their recommendations in a subsequent series of reports in 1959 in *The Lancet* medical journal, supporting and extending the recommendations of the 'yellow peril'. It is from these recommendations that today's parameters of time and temperature exposure to steam to sterilize surgical instruments is derived. A further major wakeup call happened in the early 1970s with the failure to sterilize solutions of dextrose used for intravenous infusion, resulting in the deaths of at least four patients, the so called Devonport incident.

The impact of the appearance of vCJD on surgical instrument reprocessing

Following the recognition of vCJD in the mid-1990s the Department of Health commissioned some studies into investigating the current state of surgical instrument reprocessing in the UK. Interestingly, these studies revealed remarkable similarities to the observations from the first report in 1958, a gap of nearly 40 years! Eventually the results of these studies surfaced into the public domain assisted by a whistle blower Mr David Hurrell.

In Scotland, an investigation into the decontamination of surgical instruments was commissioned and published in 2000 as the 'Old' report named after Professor David Old who chaired this group. Around this time the Department of Health published their risk assessment for transmission of vCJD via surgical instruments stating that 'on a per operation basis, risks of vCJD transmission appear strongly concentrated around central nervous system and posterior eye procedures'. All these reports were grouped together into an action plan and financing to improve the decontamination of surgical instruments across all surgical specialties.

Again its is important to put these risks into perspective and examine the numbers of surgical instruments reprocessed. In Scotland the numbers of instruments being handled at this time comprised (approximately) 37 million instruments in hospitals and 196 million outside of hospitals (general practice). The largest number of instruments being reprocessed are dental instruments, accounting for some 160 million of those reprocessed in general practice (NHS HDL 2001 (66)).

Improving the service and reducing the risk

In Scotland the task of co-ordinating and improving the service was undertaken by the 'Glennie' group, a multi-disciplinary team chaired by John Glennie, which I sat on from the outset until its closure in 2013. This group undertook further investigations regarding the practice of instrument decontamination and made more recommendations that prioritized improvement action for high risk instruments (such as those used in neurosurgery and the back of the eye) and divided recommendations into equipment, facilities, staff and management. In summary this led to the closing down and amalgamation of some hospital sterile service departments and new larger modern units being built. Activity in general medical practice was switched to either single use instrument packs or transported to hospital sterile supply departments for decontamination.

Dental instrument decontamination

Whilst much work has been on-going reducing the risk from surgical instruments used in high risk procedures for possible transmission of vCJD, there were a smaller number of studies being undertaken by my group in collaboration with a number of different colleagues to demonstrate that some types of dental instruments – such as matrix bands (used to hold large fillings in place during packing of amalgam) and root canal files – were incredibly difficult to get clean using the processes available in dental practice. These devices were subsequently mandated to single use (and still are in

Professor Smith has a Personal Chair in Bacteriology from the University of Glasgow and is a consultant microbiologist based in NHS Greater Glasgow and Clyde. Within Glasgow he is the microbiology consultant lead providing advice and leadership for the microbiological aspects of cleaning and sterilization of surgical devices ranging from dental, endoscopic and neurosurgical specialties.

Scotland!) Further work with colleagues in the then Health Protection Agency, Porton Down, helped demonstrate that oral tissues in a mouse model of prion disease were capable (under worse case scenarios) of transmission of infectivity via the oral cavity. This data was subsequently used to inform Department of Health risk assessments for the risk of transmission of vCJD via dentistry which concluded that a wide range of transmission scenarios was possible and high standards of instrument decontamination were necessary to reduce the risk. More recent investigations in collaboration with colleagues at the CJD surveillance Unit in Edinburgh, undertook a large case-control study to investigate any potential links with dental treatment in vCJD cases, but no convincing evidence was noted.

Current research initiatives

This includes using some standard and novel protein detection methodologies to investigate the cleanliness of neurosurgical instruments with the aim of using traditional quality control methods to improve the quality of cleaning and is funded by the Scottish Infection Research Network. Similar methodology is also being used to investigate the effectiveness of small instrument washers used in dental practice funded by the Chief Dental Officer for Scotland.

Conclusion

Despite the lack of any recent evidence of iatrogenic transmission of CJD via contaminated surgical instruments, significant improvements have been made in the cleaning and disinfection of all different types of surgical instruments ranging from dental devices, endoscopes and those used in neurosurgery. The overall risk reduction strategy is to work towards a higher quality of cleaning and sterilization for all instruments and not just focus on those used on high risk tissues.

EDITOR We would be pleased to hear from any reader who has had difficulty with their dentist, or any other health professional, as a result of their CJD status. Please contact Gillian Turner either by email gturner@cjdsupport.net or by phone on 01630673973.

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	85	3	7	3	98
2011	91	4	14	5	114
2012	93	5	11	0	109
2013	107	2	8	1	118
2014	90	3	10	0	103
2015*	30	0	3	0	40

* As at 6 July 2015

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

The European surveillance system for Creutzfeldt-Jakob disease (EuroCJD)

Professor Bob Will

The collaboration between national surveillance systems for CJD in Europe started in 1993 and has since been coordinated by the National CJD Research and Surveillance Unit (NCJDRSU) in Edinburgh in conjunction with the Dutch CJD surveillance system, based at Erasmus University Hospital in Rotterdam.

The initial countries involved were France, Germany, Italy, the Netherlands, Slovakia and the UK as these countries had already set up surveillance for CJD, but over subsequent years the great majority of EU countries have joined the system and collaboration has also been established with CJD surveillance groups in Argentina, Australia, Canada, China, Israel, Japan, Mexico, Taiwan and the USA.

The initial aim of the EuroCJD group was to identify any change in CJD that might be linked to the occurrence of bovine spongiform encephalopathy (BSE or mad cow disease) in cattle populations and the first task was to harmonise methods of diagnosis and case classification to ensure that there was consistency between countries in the way data was gathered.

In 1996 a new form of CJD, variant CJD, was identified in the UK and it was proposed that this new disease might be cause by BSE. One finding that strongly favoured this theory was that it was possible to establish through the EuroCJD system that no similar cases were occurring, at that time, in other European countries with small or absent BSE outbreaks. There is now very good evidence that variant CJD is indeed caused by BSE and through the EuroCJD system and international collaborations it has been possible to provide real-time data on the occurrence of variant CJD worldwide. Table 1 below shows current information on variant CJD.

Table 1 Current data on variant CJD worldwide

COUNTRY	TOTAL NUMBER OF PRIMARY CASES (NUMBER ALIVE)	TOTAL NUMBER OF SECONDARY CASES: BLOOD TRANSFUSION (NUMBER ALIVE)	RESIDENCE IN UK > 6 MONTHS DURING PERIOD 1980-1996
UK	174 (0)	3 (0)	177
France	27 (0)	-	1
R of Ireland	4 (0)	-	2
Italy	2 (0)	-	0
USA	4† (0)	-	2
Canada	2 (0)	-	1
Saudi Arabia	1 (0)	-	0
Japan	1* (0)	-	0
Netherlands	3 (0)	-	0
Portugal	2 (0)	-	0
Spain	5 (0)	-	0
Taiwan	1 (0)	-	1

† The third US patient with vCJD was born and raised in Saudi Arabia and has lived permanently in the United States since late 2005. According to the US case-report, the patient was most likely infected as a child when living in Saudi Arabia.

* The case from Japan had resided in the UK for 24 days in the period 1980-1996.

One aim of the EuroCJD system from the start of the study has been to gather and share information on this rare disease in order to accumulate enough information to allow valid analyses, which would not be possible by individual countries. This has allowed a series of publications on a range of issues, including the distribution of CJD in Europe, the frequency of subtypes such as genetic and iatrogenic cases, the factors that influence the clinical features of the illness and risk factors for the development of disease. Importantly no consistent risk factors for the development of sporadic CJD have been identified, including a recent analysis of occupation, which did not show an increased risk to medical or paramedical staff, observations consistent with the hypothesis that sporadic CJD is a spontaneous disease with no external source of infection.

Mortality rates per million population for sporadic CJD are shown in Table 2 and indicate that overall there is a remarkable consistency in the rates per country identified through the EuroCJD system, despite significant differences in health care systems between countries. There are some countries with low rates but these are likely related to limited resources or restricted access to specialist investigations and some countries with small populations (eg Iceland or Cyprus) would be expected to identify a

The development and validation of a new diagnostic test for CJD has been an important objective of the EuroCJD system

case only every few years. The gaps in information from some countries in the early years of the project reflect the time that CJD surveillance was started in each country, while the absence of data from some countries in recent years is linked to limitations in resources for CJD surveillance, a trend that may continue with the decline in public health concerns about these diseases.

The development and validation of a new diagnostic test for CJD has been an important objective of the EuroCJD system and combining data from many countries allows tests to be assessed more quickly than by single countries. The 14-3-3 cerebrospinal fluid immunoassay was first developed in the USA, but was introduced into the diagnostic criteria for sporadic CJD following work by the EuroCJD group. Publications from the group have also led to the inclusion of MRI brain scan findings into the diagnostic criteria for sporadic CJD and it is possible that collaborative research will lead to sufficient evidence to justify the inclusion of cerebrospinal fluid Real-Time Quaking Induced Conversion (RT-QuIC) in the criteria. This is a potentially important advance as this test may be specific for sporadic CJD, unlike the 14-3-3 and MRI, and allow a firm diagnosis in life.

The EuroCJD system continues to provide important data on CJD, including variant CJD, and was highlighted in a recent review of research in Europe over the past 30 years (*Horizon* magazine EU) as an example of scientific collaboration between countries addressing an issue of significant importance for public health. The participants in the project have been grateful for funding from the EU, including via DG research, DG Sanco and the European Centre for Disease Control and Prevention (ECDC) and we hope that the EuroCJD system will continue, despite the overall decline in funding for these diseases in recent years.

Table 2 Mortality rates per million population for sporadic CJD

Country	1993	1994	1995	1996	1997	1998	1999	2000	2001	2002	2003	2004	2005	2006	2007	2008	2009	2010	2011	2012	2013	Mean Rate
Australia	0.96	0.56	1.06	1.37	1.09	1.34	1.33	1.47	1.04	0.92	1.12	0.85	1.29	1.71	1.25	1.46	1.34	1.27	1.57	1.1	0.52	1.17
Austria	0.77	1.15	1.15	1.15	0.77	1.03	0.77	1.12	1.12	0.87	1.86	0.99	1.74	2.23	2.23	0.99	1.98	0.75	2.13	2.88	1.88	1.41
Belgium	-	-	-	-	0.97	1.37	1.27	0.88	1.46	2.02	1.83	1.92	1.24	1.52	1.51	0.56	0.93	1.11	1.19	0.64	0.45	1.28
Canada	-	0.07	0.1	0.44	0.53	0.73	0.89	1.04	0.87	0.99	0.85	1.31	1.3	1.2	1.06	1.44	1.42	1.03	1.35	1.74	1.05	1.17
Cyprus	-	-	1.48	0	0	0	0	0	1.43	0	1.39	1.37	0	1.30	1.28	3.84	0	0	0	1.16	0	0.7
Czech Republic	-	-	-	-	-	-	-	0.19	0.78	0.59	0.78	1.08	0.98	0.97	1.07	1.05	1.43	1.23	1.68	1.14	1.52	1
Denmark	-	-	-	-	2.09	0.94	1.69	0.94	1.12	1.68	1.86	1.3	0.74	1.47	1.1	1.1	2.36	1.08	1.26	1.79	2.32	1.46
Estonia	-	-	-	-	-	-	-	-	-	-	-	0	0.74	0	0	0	0	0.75	0	0	0.75	0.22
Finland	-	-	-	-	1.92	0.96	1.35	0.77	1.73	1.92	0.96	1.15	2.5	0.77	0.72	2.5	1.13	1.31	1.68	1.07	2.2	1.45
France	0.59	0.76	1	1.14	1.34	1.35	1.53	1.45	1.79	1.74	1.75	1.57	1.31	1.96	2.17	1.65	1.77	2.34	1.7	1.61	1.21	1.51
Germany	0.44	0.85	1	0.93	1.3	1.4	1.25	1.36	1.6	1.3	1.1	1.5	1.4	1.5	1.5	1.5	1.1	1.3	1.33	1.1	n/a	1.24
Greece	-	-	-	-	0.55	0.37	0.64	0.83	0.73	1	0.55	0.64	0.46	0.55	0.63	0.54	n/a	n/a	n/a	n/a	n/a	0.62
Hungary	-	-	-	-	1.1	0.3	0.5	0.9	1.7	0.7	0.8	0.8	0.8	1.2	1.3	1.4	0.7	1.4	0.8	1.9	1.6	1.05
Iceland	-	-	-	-	3.7	0	0	0	0	0	0	0	0	0	3.23	0	0	0	n/a	n/a	n/a	0.53
Ireland	-	-	-	0.55	0.55	1.62	0.27	0.79	1.3	1.28	0.5	1.73	1.21	0.94	0.71	1.18	0.47	0.24	1.31	1.31	1.53	0.97
Israel	0.37	0.19	0.56	0.49	0.66	0.82	0.49	0.49	0.6	0.45	1.2	1.2	0.9	0.68	0.68	1.1	1.18	1.05	1.05	0.13	n/a	0.71
Italy	0.47	0.58	0.49	0.89	0.83	1.11	1.28	1.04	1.51	1.32	1.37	1.33	1.85	1.62	1.64	1.54	1.78	1.88	2	1.8	1.5	1.33
Latvia	-	-	-	-	-	-	-	-	-	0	0	0	0	0	0.43	0.43	0	0.89	0.45	0.49	n/a	0.22
Netherlands	0.79	1.17	0.52	0.9	1.2	1.13	1.24	0.63	0.88	1.13	0.75	1.22	1.22	1.34	0.92	0.98	0.67	1.69	2.04	1.55	1.54	1.12
Norway	-	-	-	-	1.37	0.45	0.45	0.89	1.33	0.66	1.32	1.75	0.87	0.86	1.07	0.85	1.04	0.41	0.81	0.8	1.38	0.96
Poland	-	-	-	-	-	-	-	-	-	0.16	0.31	0.52	0.36	0.13	0.08	0.26	0.86	0.08	0.68	n/a	n/a	0.34
Portugal	-	-	-	-	0.5	0.6	0.5	0.5	1.6	0.6	1.05	0.57	0.95	0.48	0.76	0.38	0.57	0.19	1.23	1.14	n/a	0.73
Slovakia	0.4	0.4	0.4	0.4	0.6	0.6	0.2	0.4	0.4	1.13	0.4	1.17	1.34	0.57	0.75	1.13	0.92	0.55	1.1	0.55	0.73	0.67
Slovenia	0	0.5	1	0	0	0.5	1.5	0.5	2	2.5	1	1	1	3.5	1	1.5	1	0.5	0.5	3	2	1.23
Spain	0.54	0.43	0.51	0.69	0.79	1.57	1.27	1.14	1.57	1.36	1.44	1.36	1.81	1.53	1.76	1.37	1.4	1.28	1.26	1.06	1.08	1.2
Sweden	-	-	-	-	1.13	1.58	1.24	1.58	1.35	1.34	1.23	1.66	1.44	1.76	1.2	1.4	1.18	1.49	1.58	1.47	n/a	1.41
Switzerland	1.15	1.42	1.42	1.41	1.41	1.12	1.26	1.53	2.62	2.46	2.31	2.16	1.34	1.73	2.11	1.82	2.31	0.64	1.89	1.87	2.09	1.72
UK	0.62	0.93	0.6	0.69	1.01	1.09	1.06	0.85	0.98	1.23	1.33	0.84	1.11	1.12	1.05	1.4	1.29	1.35	1.42	1.46	1.59	1.1

CJD Support Network

Management Committee 2015

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

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

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

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

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

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

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

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

Roger Tomkins
Roger's daughter Clare died of vCJD.

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

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

Gillian Turner
CJD Support Network co-ordinator

Time to say goodbye

Derrick Biggs

After working with the CJD Support Network over the last 17 years it is time to step down. I have retired from my position with Cambridgeshire County Council and also my role of working with the Association of Directors of Adult Social Services (ADASS). It is working with the Disabilities Network of ADASS that I agreed to become its link worker with the CJD Support network.

During my association with the network I have written the Good Practice Guidelines for Social Work Professionals which was also updated on one occasion. This followed concerns that many professionals were not aware of the specific issues surrounding people with CJD and their family carers. Also many professionals were also asking for guidance due to the rarity of the condition and not having any peers to learn of their knowledge. Thus started my 17 year association.

I hope during that time I have made a useful contribution having become a member of the committee, attended many family days, spoken at conferences and received numerous phone calls from family carers and social work professionals eager for me to answer questions around social work involvement.

Having now left the professional social work scene my up to date knowledge will soon become out of date and so I felt the need for some one else to step into my position.

I have enjoyed my time immensely with the Support network. That stems from a committed co-ordinator in Gill Turner and a committed and knowledgeable committee well led first by Angus and now Richard. I believe that one of its strengths is that over half the committee is made up of family carers who have been through the trauma of losing someone close to CJD and they therefore can talk with authority.

I wish the network well in the future and will remember with fondness my time with you all.

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.

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

CJD SUPPORT NETWORK
Contact: Gillian Turner, CJD Support Network, PO Box 346, Market Drayton TF9 4WN
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