



CJD SUPPORT GROUP NETWORK NEWSLETTER MARCH 2007

www.cjdsupport.org.au

POSTAL ADDRESS

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DEPARTMENT OF HEALTH & AGEING

General information for human
pituitary hormone recipients.

National Toll Free 1800 802 306

CJD SUPPORT GROUP NETWORK (CJDSGN)

It is encouraging for us all that yet another year has passed without any further cases of CJD amongst the human pituitary hormone (hPH) recipient community. It is now 16 years and although no-one can give us an all clear I think it is reasonable that we can feel a little less concerned.

Unfortunately as we are all ageing and facing more medical procedures we are also facing screening questionnaires on admission to hospitals, in particularly private hospitals and eye and dental clinics. These questionnaires usually ask if you have received hPH prior to 1985 or if you have had a first degree relative die of CJD. It is our understanding that most of the state health departments are trying to introduce a standard set of questionnaires, and ideally questions will only be asked when a procedure involves high infectivity tissue. It is our hope that when a screening questionnaire relating to CJD is used, and the patient ticks the "yes" box, the facility has the correct procedures in place so no patient suffers delay or discrimination.

We have now established contacts in all state health departments and have people who are informed and willing to assist with any infection control issues facing our members. Experience has proved that it is better to have the situation sorted prior to admission with your doctor and the hospital/clinic so there are no last minute surprises. Too often "at risk of CJD" patients has been ready for surgery only to be sent home. If you have any concerns or any questions relating to a procedure you may require, we are here to assist you. A direct link to the infection control guidelines for CJD is available on our website.

It was the decision of the Management Committee of the CJDSGN to delay the publication of our newsletter so that time and resources could be spent producing our brochure "What is CJD?". A copy is enclosed and extra copies can be requested or you can download copies from our website. It is hoped the brochure may dispel the many myths and great ignorance regarding CJD in general. We have also produced a series of brochures to assist families affected by CJD.

We thank the Department of Health and Ageing for allowing us a budget extension to cover support for families who have lost a loved one to CJD. The number of CJD cases in Australia is consistent with the rest of the world with a little over 1 case per million of population per year. This accounts for approximately two three cases a year, most of these being sporadic with no known cause. The support we have been able to offer to these families has been very much appreciated and I would like to personally thank Mandy Newton, our family representative, for the wonderful job she is doing. It has also been encouraging to see at meetings how generous and supportive hPH recipients have been in welcoming Mandy and family members.

Suzanne

AUSTRALIAN DVD ON CJD

The CDSGN is producing an educational DVD aimed at raising awareness of CJD and providing important information to medical staff that we hope may dispel the many myths and great ignorance regarding CJD in general, as well as assist them when dealing with infection control issues.

We acknowledge and thank our friends at the CJD Foundation USA for the technical and expert content of their DVD "Confronting CJD & other Prion Disorders" that they have generously agreed can form part of our Australian DVD.

The Department of Health & Ageing has allocated funding for this purpose and we hope in the second part of this year to be able to use our DVD for educational seminars in hospitals as well as working with state health departments to provide education workshops.

We held a few seminars in several states when we were lucky enough to have Margaret Leitch, the UK CJD Care Co-ordinator, in Australia during her visit for our fundraising dinner. Margaret's seminars were extremely well attended and a great success and we are confident that we can get the same level of interest from our own seminars.

We would be very interested in the stories of those of you who have encountered difficulties and discrimination in the medical and dental system as a result of your high risk status (for familial CJD families) and low risk status (human pituitary hormone recipients). We also would like to hear the stories and experiences from our families who have lost a loved one to CJD if you are willing to be interviewed for our DVD.

We know that many of you are very guarded about CJD and being interviewed might be rather confronting. Please understand that the DVD is purely for educational purposes for use in medical seminars and not for public viewing. Please be assured that there is no pressure on anyone to assist but we do hope that you will consider our request if you feel you have something to contribute. Holding education seminars can only benefit all families whose lives are affected in any way by CJD.

Please contact Suzanne Solvyns or Mandy Newton if you would like to help.

THANKS TO A WONDERFUL FRIEND

It is with sadness that we advise members that Carol Wilson has recently resigned as a member of the CJDSGN committee and as a state coordinator. Carol was one of the first recipients of human pituitary hormones (hPH) to have her circumstances aired publicly when she appeared on the current affairs program "Hinch" in 1992.

Carol was part of the group that helped set up the Australia wide CJD support group we know today. Carol was Tasmanian coordinator for about 12 years and for the last four years she was also the coordinator for Victoria and South Australia. Since the restructuring of the support group several years ago, Carol has worked with Suzanne Solvyns as co-director of CJDSGN.

On behalf of the support group, Carol attended meetings in Washington DC in 2005 and 2006, where she and Suzanne were guest speakers at the CJD Foundation USA family conferences.

Carol's contribution to the welfare of the recipient community has been outstanding. She gave evidence before both the Allars and Senate Inquiries. In her role as state coordinator she has helped recipients through the "one-off payment" process, and has been involved in meetings with both State Health departments and Blood banks. She was involved with the Royal Melbourne Hospital incident and has been an advocate for recipients when all manner of concerns arose.

On behalf of all members of the CJDSGN, the management committee would like to thank Carol for the wonderful contribution she has made in so many areas and for the compassion and help she has given to people who sought her assistance. Throughout her time in the Support Group, Carol has always had the love and support of her husband Lex and daughter Lisa. We wish her well as she moves on to another phase of her life and the opportunity to spend more time with her family.

Suzanne, David and Mandy.

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BONDED AGAINST CJD

GALA FUNDRAISING DINNER A HUGE SUCCESS

We are delighted to report that our gala fundraising dinner held on November 10 in honour of International CJD Awareness Day (November 12) was a huge success.

We must thank our MC, **Geoff Cowan**, of TVSN who generously donated his time to support our cause and did a wonderful job. Thanks is also due to our guest speaker **Jennifer Cooke**, journalist and author of *Cannibals Cows & the CJD Catastrophe* who has been a tremendous spokesperson and advocate of the CJD SGN for many years and we are extremely grateful for her support and participation at the event.

Our guests, who included our special guests **Margaret Leitch**, CJD Care Coordinator (UK), **Steve Collins** and **Alison Boyd** from Australian National CJD Registry, were all treated to an evening of fine food and fantastic entertainment courtesy of **Total Gravity**, a wonderfully entertaining and charismatic covers band who kept the dance floor pumping well into the night.

We would also like to thank our guest auctioneers for the evening, **Bruce Newey** and **Pat Nati** for such an entertaining auction, **Look Print**, for donating the display banners, **Nadine Solvyns** who was creative designer of our program and **Emerald Press** who donated the printing of all the programs and donation envelopes. We are eternally grateful to you all.

The funds raised surpassed all of our expectations and we couldn't have been more pleased with the result. We would also like to thank all of you who attended and contributed to the evening in many, many ways and thank those of you who made such generous donations. A big thanks to our helpers on the night who did a fabulous job - **Cameron Howe**, **Karina Ralston** and **Lauren Ralston**.

A special note needs to go to our friends and wonderful sponsors who organised or donated the many amazing items for our Live and Silent Auctions:

Air New Zealand, AJC, Alphapharm, Australian Rugby Union, Bianca Velder, Lisa & Pete Bayliss, Book & Music Market, Ruth Brown, Canon, Ceroc & Modern Jive Dance Company, Pierre Colin-Thome, Tova Ducker, Geoff Dunne, Leanne Geraghty, Alison King, La Prairie, LG, Margaret Leitch, Dr Clement Loy, Nerida McLane, Susie Meehan, Nati Bros Roses, Neway Transport, News Limited, Graham Philp, Sarah Ridgeway, South Pacific Lancaster Fragrances, Sydney Convention and Exhibition Centre, Sydney Seafood School, Sydney BridgeClimb, Kate Thompson, TVSN, Unique Stay Tasmania (Solvyns family), Jennifer Vernon, Video Ezy, Warraroong Estate Winery, WizKids (Osborne family), Carol and Lex Wilson and Dave Wong.

Such success has inspired us to hold the event on an annual basis and we hope that you are able to join us this coming November for what is certain to be another great success.

Suzanne and Mandy.



ADDRESS TO the INAUGURAL CJD FUNDRAISING DINNER

“Celebrating CJD Awareness Day” Friday November 10, 2006.

Guest speaker: Jennifer Cooke

Author of ‘Cannibals, Cows & the CJD Catastrophe’

Good evening everyone,

Welcome to the inaugural CJD fundraising dinner.

It’s fantastic to see you all here – and the place is full!

Well done Suzanne Solvyns and Mandy Newton in getting you all here.

As I look around and see everyone here, there are some very familiar faces. Some are vaguely familiar and some – of course – are getting an introduction to Creutzfeldt-Jakob disease and its far-reaching ramifications for the very first time tonight by coming to support a good cause.

My introduction to this awful disease came almost 14 years ago to the day. Amazing and tumultuous things have happened since then – far too many to detail here.

Back then I was only several months returned from nearly four years working in London and Hong Kong – mainly as a crime and legal reporter.

So it was quite a shock to come back and find myself suddenly the Sydney Morning Herald’s medical writer. It was even more of a shock when the phone rang one Thursday afternoon in November 1992 at the old Fairfax Headquarters in Broadway (the locals will know it as one of the ugliest buildings in the country).

On the phone was a stranger – a Dr Lynette Dumble – who would loom large on my horizon for some years to come. She was a research scientist from the University of Melbourne and she had rung up cold to tell me that a paper she was to present the next day at a bioethics conference would warn of the potential for a world-wide epidemic of Creutzfeldt-Jakob disease. I had to ask her to spell it several times and wished for the first of many a time why some neuropsychiatrists called Smith and Jones hadn’t reported before it Hans Creutzfeldt and Alfons Jakob back in the 1920s.

The story I wrote about Dr Dumble’s dire warning was splashed across the front page the next day. It was scary stuff – how human hormone drug recipients may unwittingly pass on their risk of getting CJD from long-ago injections to others – through blood, organ and tissue donations.

Thereafter my phone rang hot – not just for days, or even months.

It lasted years. It’s still happening.

Luckily, Dr Dumble’s scenario did not come to pass. But it was a taste of what was to come – what now looms – that could be worse. That is the spread of BSE-related variant CJD among humans via blood products and surgical instruments from people who are incubating the disease without symptoms.

As I stand here 14 years after that fateful phone call I see some of the original members of the old CJD Support Group Inc. I see Geraldine Brodrick, the long-term Queensland co-ordinator and one of the most famous recipients of human hormone therapy in the world. And of course many of you know Suzanne Solvyns, who has been part of the support group community almost from the outset, the co-ordinator in NSW for years and now, with Carol Wilson, from Tasmania and unfortunately too ill to attend tonight, the inaugural co-chair of the International CJD Support Alliance formed in July this year.

She’s a bit of a powerhouse, the old Suzanne. She pushed and pushed until she persuaded me to attend the American CJD Foundation Family conference held in Washington DC this year – at my own expense. So I took my husband, as it was the first “holiday” we had ever taken alone (without children) since we got together 10 years ago.

It was well worth it. My husband asked knowledgeable questions of several scientists – as he ought to after a decade of listening to me. And I was honoured to meet some of the people who had lost relatives to this awful disease and gain more insight into how CJD affects families.

Professor Robert Will, one of the stars of my book *Cannibals, Cows & the CJD Catastrophe* and one of the giants of CJD research who tracks CJD cases worldwide, was shocked to see me in the US. It was a full 10 years after we had first met over some brain slides at the Creutzfeldt-Jakob disease Surveillance Centre in Edinburgh.

“Goodness,” he said, recognising the Australian accent. “Are you STILL following this?” Yes, I replied, thinking it a bit rich that HE should be asking ME when his first big paper on the subject was published in 1979 (the year I started in journalism).

Dr Will’s comment reminded me of the aftermath of March 20, 1996 – the day the British Government finally admitted a link between bovine spongiform encephalopathy and variant Creutzfeldt-Jakob disease.

That meant the size of my proposed book – which had only been accepted for publication the day before – had suddenly blown out. I was already intending to skim lightly over neuropathology, gynaecology, endocrinology, virology, pathology, veterinary science and cannibalism, in between terrible losses to families around the world.

After the cows came home, so to speak, I had to include everything known until then about sheep and mink and deer and elk and goats and hamsters and macaque monkeys and chimpanzees – any animals really, that had ever been experimentally inoculated with a spongiform encephalopathy.

The book length blew out from an expected 90,000 words to 136,000 (I don’t think that included references). But hey, it won the 1999 Eureka Science Book Prize, the premier book prize in Australia for communicating science to the general public, so the publishers stopped moaning.

When the first mad cow was discovered in Canada, then in the US, then Japan, I got more phone calls – even though I was in a job that did not involve writing. I spent Christmas Day 2003 dripping in ferocious heat in my father’s unairconditioned study writing another page one story as my children waited with their cousins to open their gifts.

And the calls come still: every time there is a scare with potentially contaminated instruments at a hospital; every time a new case of variant CJD in a blood donor in Britain is announced; and, when Australian human hormone recipients are discriminated against by health professionals who are ignorant of CJD itself, let alone their low risk of developing it one day.

Watching the CJD Support Group grow (during which I was sometimes used as a sounding board between factions in the early days) has been nothing short of fascinating. From two embryonic cells, it fused into a cluster and then a solid, angry but focused mass with enough relentless energy and clout to force the establishment of a Senate Inquiry into what it considered was an unfair compensation settlement to those who had been suing the federal Government.

As the Support Group Network grew – and remember it was the first of its type in the world – it continued to be funded ostensibly for the hormone recipients who are at risk of iatrogenic or medically transmitted CJD. But it also provided support for anyone who sought help for ANY type of CJD – like sporadic CJD or familial CJD.

We have yet to see a case of variant CJD in this country but the experts agree it is inevitable given our close migration and travel links with Britain particularly.

This is quite apart from the extremely rare familial offshoots including Gerstmann-Straussler-Scheinker syndrome and Fatal Familial Insomnia (which is the subject of a new book just out in the US).

Last year Mandy Newton – who with her sisters bravely went public with their close link to familial CJD – became the driving force behind a new arm of the Australian CJD Support Group Network. She is now its family representative. The new arm of the group has evolved thanks to continued funding by the federal Government and helps support the families of CJD sufferers, particularly those with a familial prion disease.

I think the only help that has not been given by Suzanne Solvyns, Carol Wilson, Mandy Newton and other former hard-working members of the support group and its inter-Governmental committees over the past 14 years is to the relatives of sufferers of kuru. And that’s only because they are in the remote highlands of Papua New Guinea and can’t call the toll free number! It’s been wonderful to watch the group achieve great things that seemed so unattainable a decade and a half ago.

Who'd have thought that the Allars Inquiry, set up in 1993, would have revealed so many suspected but unpalatable truths so soon after the first calls for an inquiry into the Australian Human Pituitary Hormone Program.

Who'd have thought that a legal test case against the Commonwealth Government, also instituted in 1993, could have pointed the way – even though it ultimately settled in 1997 – to a successful outcome for similar litigation in England's High Court. The mere fact that the test case was allowed to proceed past the first challenge by the Commonwealth showed that the law was prepared to shift in favour of people who developed a psychiatric injury through anxiety caused by the mere communication of bad news. Bad news that changed lives overnight, ended others, ruined marriages and friendships and introduced stigma where it had never been before – all because they were told they may contract a fatal disease from a Government-sponsored, elective medical program that promised to correct infertility in adults and short stature in children.

But as long as these milestones continue to be passed, the phone calls will continue. And I will no doubt answer them. I have already based one university thesis on the CJD test case in Australia. Next it'll be the PhD!

After 14 years even my friends and family have stopped saying "how is that Schwartzkopf thing going"? Now they just ask how the mad cow is.

One thing seems certain from where I stand. Involvement with anyone fighting for more research, more help, possibly a drug that might actually treat CJD in the future has to be ongoing.

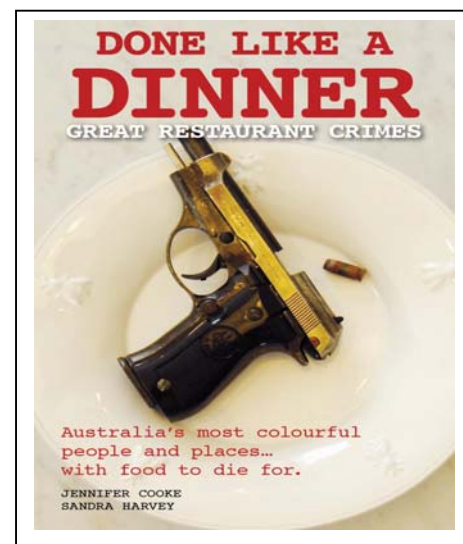
It's a bit like the Hotel California. You can check in _____ but you can never leave.
So dig deep tonight and hopefully see you all here next year!
Jennifer Cooke

Jennifer's latest book is 'Done Like A Dinner' - a true crime book that looks at crime with a strong restaurant link. Not only just the food that's criminal! And it comes with recipes ... Park Street Press, available in book stores or direct from www.media21publishing.com.au

Here is a recent review

The Weekend Australian SAT FEB 10, 2007, Page 010

CRIME FILE By *GRAEME BLUNDELL*



FOOD is to the mystery story what Holmes is to Watson: an inseparable best friend. It was never safe to eat in the classic British golden-age thriller, the main course always served with poison on the side. And the American PI thriller usually involves as many meals as murders.

As Jennifer Cooke and Sandra Harvey's entertaining *Done Like a Dinner* (Park Street Press, \$27.95, 208pp) reveals, shootings, stabbings, gangland assassinations, stings, bribes, blackmail and kneecappings happen around food in real life, too. Resourceful, intrepid and convivial, Cooke and Harvey have wittily collated some of the best-known local criminal events in this cook's tour of crooks.

Among them: the mysterious shooting of Ray "Ducky" O'Connor in Sydney's Latin Quarter in 1967 amid the oysters kilpatrick, cocaine and champagne cocktails; the 1978 late-night siege at Melbourne's "Spaghetti Speakeasy", the Italian Waiters' Club; the arrest of drug baron George Savvas in 1997 at Sydney's most expensive Japanese restaurant. They use witness accounts, public records, police contacts and "the odd waiter or two" to chart their gastronomic underworld.

The fact that these authors can turn true crime into popular entertainment says a lot about the way art mirrors the world, but also filters it into something, well, digestible.

INFECTION CONTROL GUIDELINES

The Australian 'Infection Control Guidelines for the Prevention of Transmission of Infectious Diseases in the Health Care Setting' provide a guide to all health care professionals including doctors, nurses, dentists, and nursing home staff. In the current edition of the guideline (2004), there is a specific chapter on CJD (Chapter 31) with additional information on CJD in Appendix 9. The CJD sections are currently under review by the Communicable Disease Network Australia with an aim to provide a more concise and easy to follow single chapter. The new chapter will have tables and charts to allow a health care professional to quickly decide on the level of infection control required for each procedure and will outline what additional precautions may need to be put in place when a procedure may expose a tissue of high infectivity for CJD. This should enable the health care professional to provide the highest level of care to all patients, regardless of their risk status for CJD. The Infection Control Guidelines are available to all health care facilities and the version in use can be downloaded in PDF form from the Department on Health and Ageing website (www.health.gov.au). The CJDSGN has been consulted in the development of the new CJD chapter, along with experts in the fields of infection control, nurses, neurologists, dentists and CJD scientists. The CJDSGN will be informed once the chapter is complete and ready for public dissemination.

Tova Ducker
Assistant Director
Communicable Disease & Health Risk Policy
Office of Health Protection

Education visit by Margaret Leitch UK Care Co-ordinator

Margaret Leitch is the national care co-ordinator in the United Kingdom. She takes care of CJD patients Scotland, England Wales and Northern Ireland. Margaret was sponsored by the National CJD surveillance unit based in Edinburgh to attend our fundraising dinner. She offered to do some presentations while she was here. We managed to carry on our CJD Awareness Day campaign for a week and made a good start on our planned education programs.

This was our week. . . .

Monday: Margaret and I had a three-hour with four members of the Commonwealth Department of Health and Ageing, including the director of communicable diseases. We are hopeful we may have shown the need for our own care co-ordinator to assist patients and families dealing with CJD in Australia.

Tuesday: We presented at the Royal North Shore Hospital to a large number of doctors and nurses with the meeting extending to two and a half hours.

The CJD Support Network introduction outlined issues facing "at risk patients" accessing medical care, Margaret's presentation was aimed at educating hospitals and staff about CJD, caring for a CJD patient and her role in the UK.

Wednesday: We flew to Melbourne for a meeting at the Australian National CJD Registry attended by some members of the research teams based at the University of Melbourne who were joined by registry staff.

Thursday: We attended a meeting in Hobart with the president of the Australian Infection Control Association Inc, and discussed infection control issues for "at risk groups". Margaret did a presentation at the palliative care section of Hobart Hospital.

Friday: With a 4.30am start we caught a flight to Sydney, and joined by Mandy, we drove to Newcastle for two successful and well- attended presentations at the John Hunter Hospital.

Monday (again): Margaret made another presentation at Westmead Hospital followed by an afternoon meeting with members of the NSW Health Department - all keen to learn from Margaret's experiences.

We are planning education awareness workshops for later this year. It was encouraging to see the level of interest about CJD by so many clinicians..

Suzanne.

THE FAMILY ARM

Since it's inception two years ago, the family arm of the CJDSGN has been extremely busy hosting meetings in Melbourne, Sydney, Hobart, Adelaide & Perth. As I outlined in our previous newsletter, my volunteering as family representative of the CJDSGN was prompted by my father's death from familial CJD (fCJD) in August 2004. Since then, I have had the opportunity to meet many families and individuals who have lost loved ones to familial (fCJD) and sporadic CJD (sCJD).

I must say that this has been an extremely rewarding, if not - without exception - always highly emotional experience. To those of you with whom I have had the opportunity to meet, I would like to thank you for attending our support group meetings and sharing your stories and experiences with one another. I have always believed that the opportunity to share our stories is extremely cathartic and can be highly beneficial to "new" families who have only very recently lost someone to CJD.

One of our family arm members said something to me once, the first time we ever made contact with one another. It was in the very early days after losing my Dad and I have never forgotten her wise words. She told me that for her, the spectre of CJD has slowly receded over time and while it manages to raise its ugly little head from time to time, its impact is no longer as lasting or severe. I found great comfort in these words and two years down the track I am beginning to understand what she means. It is my hope that of our members in the family arm one day have this same realization - to acknowledge that our special loved ones were here, bringing love and happiness into our lives but without the need to dwell on the way in which they were taken from us. To put a different spin on it, I would say that I truly hope that for all of our families, the stigma, fear and confusion that comes with CJD recedes and that the knowledge they gain about CJD is as empowering for them as it has been for me.

We have a very busy year planned for 2007 with meetings for the family arm scheduled for Qld and the NT. Meetings in NSW and Victoria are also on the agenda. I encourage all our families to be as active as possible in the support group. You may have strengths and talents that may be helpful. Please, if there is something you would like to contribute don't hesitate to discuss it with us.

I look forward to meeting with you all again this year and wish you all much happiness and good health in 2007.

Mandy Newton.
Family Representative.

WARNING;

The next few pages does relate largely to families who have lost a loved one to CJD and that this may be particularly confronting to hPH recipients and others at risk of CJD.

We understand it may difficult to read about the CJD experience and its affect on the families left behind. We would encourage you to approach the content on the next few pages with thoughtful consideration, and should you choose not to read on, we ask for your respect and understanding. As members of the Australian CJD community, we believe that every one at risk of CJD, iatrogenic, sporadic and familial/inherited forms should be heard.

We have learnt from our friends at the CJD Foundation USA, CJD Support Network UK, CJD Support Network Japan and others that we all share a common goal.

The CJDSGN represents all families affected by CJD. It offers support and assistance to anyone at risk of CJD or who has lost a loved one.

The Network also encourages raising money for research so that a test, treatment and cure may become a reality.

Suzanne, Mandy and David.

MICHAEL'S STORY

"No idle chatter"

Complete strangers come together for many reasons: hobbies, working bees, sport, a long plane flight. The conversation is often idle chatter about life's shared experiences. But rarely in my experience do people come together to share such intimate details as was the case late last year. Talking about the loss of a loved one – in my case my brother – and to hear the stories of other people whose lives have been and, in some cases, continue to be affected by CJD is no idle chatter. This is human interaction at its most personal.

Why did I go? My father also died of CJD, but in the luck-of-the-draw type circumstances that seem to epitomise this illness, I was genetically tested for the familial strain and was found not to have it. As I am the only remaining member of my father's family that could have contracted the disease, CJD has become a thing of my past and, as a father of four young children, thankfully not my future.

But to watch my brother over several years go from a healthy and fit adult into a contorted, shrunken, brain-ravaged shell of a man is not something you just leave behind. I learnt and experienced a great deal during this time, and after his death it took me at least a couple of years before I took the CJD test. What dentist will fix my teeth? Should I cancel my organ donor certificate? Can I get life insurance?

One by one, explanations had to be made to various parties. In a world where it appears that CJD is just beginning to enter the medical vocabulary in a more widespread way, and because of the transmission of the disease through blood transfusions, it is not something you can just keep quiet about. And that is why I went to the meeting. It was not just the emotional "stuff", but the very practical matter of how one dealt with the world. While in a day-to-day sense I can leave CJD behind, I now feel part of a small community of people whose lives have been affected in a very dramatic way.

At the meeting, when the opportunity to speak about one's own experience was offered, a woman sitting next to me told of how she had also been tested but wished to keep her result private. She then went on to tell in a remarkably honest and open way how CJD had affected her family.

Towards the end of the meeting the same woman confessed that it was the first time she had talked about her experiences of CJD to a group of people. And she also made known the result of her test. I hope I will not upset her by writing about this, but, for me, it was an amazingly brave thing to do in front of people whom she had never met several hours before. It was certainly no idle chatter.

FUNRAISING

Two of our families are about to embark on their own fundraising campaigns, which are endorsed by the CJDSGN.

Riding for Dad

Ronnie Donnellan (Jnr) will begin his Riding for Dad fundraising campaign in May. He plans to cycle – 1000 kilometres from Alice Springs to Ulura and back. His father Ronald (Snr), lost his battle with CJD in June 2006.

Hel's Angels

In addition to Ronnie's amazing ride, the family and friends of Helen Ferry plan to take to enter the Gold Coast Half Marathon, the 10 kilometre run and a shorter walk.

They will embark on their "Hel's Angels" fundraising campaign in July. Helen lost her battle with CJD in March 2005.

We encourage you all to support Ronnie and the Ferry family. If you would like to make a donation (no amount too small, no amount too big!) contact us. Tax deductible receipts will be issued.

Please support these families and their efforts to raise awareness and money for CJD research so that other lives are not cut short by CJD.

Mandy.

"Riding For DAD"

Raising money for research into
Creutzfeldt-Jakob Disease (CJD)

Ronnie Donnellan (Jnr) will be cycling from Alice Springs to Uluru (Ayers rock) and will return to Alice to complete the last stage of his journey by riding in the Bangtail Muster Parade 7th May 2007.



Be a part of Ronnie's ambition to raise much needed funds to promote awareness and support research into CJD.

Ronald Donnellan (Snr)

11th November 1945 - 27th June 2006



Ronald was well respected in the Alice Springs Community as a true gentleman and recognised as a great mechanic. He would help anyone at any time. He was a man of few words and a devoted family man. He was extremely generous, he didn't live in a material world, what he gave people was his love and knowledge.

**All sponsorship and donations can be deposited into the CJD Support Group Network (CJDSDN) fundraising account at the Commonwealth Bank
CJD Support Group Network Pty Ltd - Account No: 06 2313 10183525
(Please ensure all automatic transfers specify "Riding for Dad")
Or call Suzanne - CJDSDN on 1800 052466 with credit card details
All donations made will be acknowledged with a tax deductible receipt.
Or contact Ronnie directly**

**Contact Ronnie Jnr: PO BOX 1719, ALICE SPRINGS, NT 0870
Telephone: (08) 8952 0439 or Mobile 0439 766 140**

No White Flags – The story of Helen Ferry a sporadic CJD Sufferer.

My Mum was a bright and bubbly 53-year-old who loved and lived life to the fullest. After raising five children, Mum began her working career late in life. She loved her job, which brought her into contact with people, her true passion. She would walk or ride her bike to work most days and return home to enjoy a meal cooked by her loving and doting husband of 33 years. Together they would share the joys and the woes of their working day before Mum would retire to the couch with her mobile phone. She would settle in for her daily ritual of chatting to one of her five children who were scattered across Australia.

In November 2004, she began to experience back pain and numbness, somewhat similar to a bulging disc. She went to a GP, a physio, a chiropractor and then back to her GP, with no improvement. By her grandson's 1st birthday on the 2nd of December, (the same day as Mum and Dad's 33rd wedding anniversary), she was preoccupied and we thought, 'over concerned' with her 'back pain'. She loved and adored her only grandchild but at his 1st birthday, refrained from picking him up for fear she might further damage her back. This didn't stop her showering him with kisses and cuddles but you could see the pain in her eyes as she watched his aunts and uncles carry him around.

Throughout December her back pain worsened and the stiffness progressed down through her bottom and into her left leg. By Christmas we all knew it was no longer simply back pain. Mum was sick. There was a considerable difference in her demeanor. The woman, who once walked 5-6km's regularly, was struggling to reach the end of our road, roughly one kilometre. Mum knew something terrible was happening to her then but we refused to let her entertain these thoughts. We were sure we would soon find the answer and she would be well again in no time.

Late in December, Mum went to Canberra for her first specialist appointment. A number of us were by her side and as always, Dad was the stronghold. Seven adults and one thirteen-month-old baby squashed into a two-bedroom unit. A long series of MRI scans and other tests ensued, including a lumbar puncture test for CJD, which returned a negative result. Terms like 'Cancer', 'Parkinson's', 'Multiple Sclerosis', and 'Motor Neurone Disease' were thrown around. With each test our hearts would sink and then rise as a negative result was returned and another horrible illness was struck off the list. We remember Dad saying, "Well the good thing is we have ruled out a lot of the worst ones." But a haze of confusion still hung around as Mum's symptoms progressed. The numbness moved up her back and down her left arm and an involuntary twitch began in her left arm. Mum was scared, confused and embarrassed as she watched her limbs take on a mind of their own. Her speech became slower. It took her longer, not to formulate her words, but to say them and she became increasingly frustrated by this. Her diet reduced and she began to lose weight. But she was still 'Mum'. Her personality never changed.

By the end of a week of tests and treatment in Canberra, with no positive results and no answers, we travelled to the Royal Prince Alfred Hospital (RPA) in Sydney where she was to be treated by a Professor of Neurology - the Head of RPA Neurology. Mum spent 10 days there where another series of tests were carried out. Mum's fears rose and her ability to sleep declined as she worried through the nights. "No white flags" became our motto as we urged her not to let this thing beat her. The involuntary muscle movement progressed to her legs and she now was unable to walk unassisted. On the January 22, Dad's birthday, we were finally given an answer to our questions, 'Cortico basal degeneration' - a slow degenerative disease of the brain. We were told that Mum had a maximum of six years to live and that she would be bedridden within two.

Later, Mum said that as early as December 2004, she would lie in bed awake at night and 'feel the disease pulsing through her veins'. She said she knew way back then that there was something seriously wrong with her. By the time they diagnosed her she was almost relieved and she could stop worrying. We, however, were shocked and devastated. The prognosis was unbearable for us. The Professor told us Mum would have been deteriorating slowly for the last 1-2 years. But we were left confused. It had been a quick but 'staged' takeover of her body, occurring over the last few months. It had moved from one limb to another in a segmented fashion whilst her mind had remained as alert as ever. The Professor told us we were wrong and probably 'hadn't noticed her deterioration before now'. But the Professor was wrong, not us.

Mum was flown home via air ambulance and Dad busied himself planning the next 2 years of his life with his wife in a wheelchair. Holidays and wheelchair access renovations. He was set to make the next 6 years as easy and as special as he could. We children also returned to Bega. We were all in shock and needed time together and time with our beautiful Mum. Major decisions were made in that time - all based on the belief we had about 2 good years left with Mum before she would be bedridden.

At our youngest sister's 21st birthday on February 8, there was not a dry eye in the room as Mum insisted on speaking. She sat in a wheelchair with her family surrounding her and my brother fighting hard to discreetly hold down Mums twitching leg.

Afterwards, our family started to return to their respective homes and to their jobs, believing we had another several years with our beautiful Mum. However Mum continued to deteriorate at the same pace, if not quicker, before our very eyes. She kept losing her balance and it now took two people to shower her. She was using a walking frame but that was quickly replaced by a fulltime wheelchair. The twitching and spasms became severe. Her arms stiffened and her hands became clenched. What was going on? The RPA had turned us away with an expert diagnosis and a prognosis of a slow deterioration over two years.

Much of our anguish was relieved after a chance visit to Bega by a neurologist, Dr Leo Davies. For the first time, we felt someone really listened to us and to Mum and took notice of Mum's rapid deterioration. He offered some possible alternatives to the disease and arranged for Mum to be flown back to RPA. He told us it was not good enough for Mum to have been discharged earlier and promised she would not be discharged again "until we got to the bottom of this".

So amid more speculation about her sickness and the possibility of a wrong diagnosis and the faint hope that this might be something else, something curable, she was flown back to RPA in Sydney. The entire family followed. It took only a few days this time. About 6pm one night Dr Davies entered Mum's room. The mood was cheery as Dad relayed a joke Mum had played on him the night before. The look on Dr Davies face stopped all conversation and made our hearts race. We children left the room leaving Mum and Dad to hear the news.

It was confirmed Mum had Creutzfeldt-Jakob Disease (CJD). Dr Davies said she had about two to three months left.

The next few weeks are a blur as Mum returned home to Bega Hospital and eventually to her own home. Her brother, a nurse, moved to Bega with us, to care for Mum. Nine adults and one small child lived under the same roof caring for our dying Mum, wife and sister. Each child took the opportunity to have private time with our Mum to care for her, hold her and comfort her as the symptoms of this horrible disease took over her body.

She was finding it increasingly hard to speak and to swallow and she had this horrible sensation that she was falling. She would call out in fright. All we could do was comfort her and be by her side every moment. Her communication lessened every day but unlike many sporadic CJD sufferers, she knew who we were right to the end and she understood everything.

On March 5, 2005, two weeks after she had been diagnosed with CJD, six weeks after being given an incorrect prognosis of six years, Helen Ferry passed away in her own home and in the arms of her husband and her 5 children. The roller coaster ride of CJD had lasted but a few months and had left a family reeling in its wake.



In memory of Helen Ferry

Hel's Angels



Raising money for research into
Creutzfeldt-Jakob Disease

If Life was a Marathon....

Helen's marathon was cut short by this debilitating disease, which is unknown to many. We, the Ferry family and friends the 'Hel's Angels' plan to take to the roads in the **Gold Coast Half Marathon** on 1 July 2007. We are pounding the pavement in order to raise much needed funds to help prevent future marathons (lives) being prematurely cut short from Creutzfeldt-Jakob Disease.

Please support us in our attempts to promote awareness and raise money for the research into CJD - the illness that took our mother, wife and friend.

Helen Ferry died in March 2005 at 53 years of age, leaving behind her husband, and 5 children.

Helen was a loving mother and dedicated friend, who would do anything for anyone. She was well known and much loved in her community and her spirit of helping others lives on.

So little is known about Creutzfeldt-Jakobs Disease (CJD) that it took agonising months in hospitals, thousands of kilometres from home, with a variety of specialists, searching for a name to the mystery illness ailing Helen. Finally she was diagnosed with CJD, a terminal brain disease with no treatment and no cure.

She came home to her family where she died two weeks later.



Donations can be made via Cash, Credit Card or by depositing directly into the following account. Please ensure all automatic transfers are specified to the Hel's Angels.

Tax deductible receipts are also available.

CJD Support Group Network Fundraising account
Commonwealth Bank BSB 062 313 Account number 10183525

For more information on CJD please go to www.cjdsupport.org.au

Family Conference in Washington DC

By Mandy Newton

Last July I was lucky enough to attend the CJD Foundation's annual family conference. It was an amazing experience to meet so many families who had also lost a loved one to both sporadic CJD (sCJD) and familial CJD (fCJD). It's difficult to put into words, actually just how beneficial and rewarding it was. On the final day of the conference after what becoming an experience that I knew I would never forget, I was invited to attend a special break-out group for familial CJD families.

I remember walking into the board room expecting to see just a handful of people, as is the usual scenario at our meetings in Australia. I was absolutely gob-smacked to see around 30 or so people sitting around the table - all from familial families. We took turns introducing ourselves and just when I thought I was recovering from my initial shock over the number of people in the room, I was further taken aback when at least 90 percent revealed that they, like me, had tested positive to the genetic mutation. It was as though there was an instant bond and while it sounds corny, there was such a sense of comfort in knowing that these people "got me". They probably understood how I felt more than anyone else could-even my closest and dearest friends who had been nothing but supportive since my genetic testing results were revealed.

The conference was excellent on a personal level, but on an entirely different level I really have to congratulate the CJD Foundation for putting together such a fantastic program. I encourage all families - familial and sporadic - to consider attending the next conference this July. We have nothing of this scale in Australia as we lose so few people to CJD and consequently have fewer families active in our support group.

If you are able to, please consider it. You can get details from the CJD Foundation website (<http://www.cjdfoundation.org/>).

We have included a photo of the CJD International Support Alliance which held its first meeting during the conference. If you look closely you'll see Suzanne, Carol and me in the shot together with our wonderful friends from the CJD Foundation, the CJD Support Group Network Japan and the CJD Alliance (UK).

NEW INTERNATIONAL ALLIANCE

The inaugural meeting of the CJD International Support Alliance took place in Washington DC in July 2006. CJDISA is co-chaired by Suzanne Solvyns, national co-ordinator of the CJD SGN, Australia and Florence Kranitz, president of the CJD Foundation, USA. It includes six support networks from 4 countries:

CJD Foundation, USA

Insight USA

CJD Support Group Network, Australia

CJD Support Network, UK

CJD Alliance, UK

CJD Support Network, Japan

It is hoped that a European family group will be established.

In the following edited extract from the Sydney Morning Herald the Health & Science Page of October 19, 2006, Jennifer Cooke was the first to report about the alliance.

"It is not every day that the welcome speech at an international scientific conference is delivered by a non-scientist with no political or research clout. Florence Kranitz brought a new dimension to a gathering of hundreds of researchers" who met in October 2006 in Italy for a major Prion disease conference. "Our primary goal is a simple one: when we see a need we do everything humanly possible to fill it," Kranitz told 900-odd delegates to the conference."

"The [CJD International Support Alliance] aims to represent people who are healthy now, but at risk of developing a prion disease as a result of medical procedures such as human hormone injections, dura mater grafts, use of contaminated surgical instruments or blood products transfusions. It covers those exposed to BSE-contaminated beef products and the elderly, who die of sporadic CJD at a rate of one per million per country per year. . . "Solvyns describes the new alliance as a broader approach to 'sharing information and resources on behalf of patients and families...the world over'."



CJD FOUNDATION USA **By Florence Kranitz**

The 4th Annual CJD Foundation Family Conference was held in Washington D.C. at the Washington Court Hotel on July 7-10, 2006. Grants from the CDC, Pall Medical, Odyssey Health Care and a record breaking attendance made this year's event our most successful.

We began with an informal reception Friday evening which offered family members and scientists the opportunity to renew old acquaintances or begin new friendships. Meeting in this way gave our families the chance to share their stories of love and loss in an informal environment. Representatives from the Centers for Disease Control and Prevention, the National Institutes of Health, the Food and Drug Administration and the Department of Defense also attended as our guests.

Saturday morning opened with a memorial service at which 52 loved ones who had died of CJD were remembered. Each name was read as a flower was placed in a vase. The vase then remained at the front of the room during the entire conference. The day continued with 10 presentations from:

Bernardino Ghetti, Professor, Indiana University, expert in GSS.
Pierluigi Gambetti, director, National Prion Disease Pathology Surveillance Centre
Robert Will, Professor of Neurology, University of Edinburgh
Neil Cashman, Brain Research Centre, University of British Columbia
Richard Knight, director, National CJD Surveillance Unit, Edinburgh
David Kocisko, NIH Laboratory of Persistent Viral Diseases
James Sejvar, neuroepidemiologist, Centres for Disease Control
Michael Geschwind, Memory and Aging Center, University of California
Gene Major, National Insitute of Neurological Disorders and Stroke
Joseph Cervia, , Pall Medical

Our annual banquet was held on Saturday evening. Tom Sheridan, president of The Sheridan Group was our guest speaker.

Sunday morning the Medical Education DVD, "Confronting CJD and Other Prion Disorders was introduced, and Tracy Kedzierski followed with a report about the CJD Foundation Family Questionnaire. Pat Lillquist, Ph.D., of the New York State Department of Health reported on the surveillance and education concerning CJD that is carried out in New York State. Deana Simpson, RN, founder and director of CJD Insight offered an excellent presentation of the history of CJD and encouragement for those struggling with the reality of CJD.

Don Simms of Belfast from Northern Ireland, father of Jonathan, shared the powerful story of the journey his family has taken over the last five years. His DVD presentation outlined the facts in a dramatic rendition of his fight for the right to use pentosan polysulphate (PPS) as a treatment for his then 17 year old son diagnosed with variant CJD (vCJD). Jonathan is still alive five years after treatment began making him the longest living variant CJD (vCJD) patient. A father's commitment and a son's struggle for life were vividly presented.

During lunch on Sunday there was a special meeting for family members coping with familial CJD. About 30 people attended and were encouraged to share their stories, concerns, questions and hopes. Deana Simpson was moderator and Dr. Gambetti attended as advisor to this group. At the request of all who attended, it is planned to include this event again next year. From this initial meeting it was decided that Deana would host an online support group twice a month for those affected by fCJD.

Our friends from "down under" were, as always, a welcome addition to the conference. Suzanne Solvyns, Carol Wilson and Mandy Newton offered a report on the activities of the CJD Support Group Network in Australia. They are growing considerably and are conducting more meetings in each state of Australia as well as hosting fund raising events.

Dr Muneto Ueda, chairman of the CJD Support Network in Japan attended this year's conference for the first time and brought a group of four colleagues with him. Dr. Ueda's very comprehensive and moving presentation focused on cases of iatrogenic CJD. In Japan the risk of iatrogenic transmission from CJD- contaminated dura mater transplants is 1 in 100,000.

Putative Treatments

Graham Steel
Information Resource Manage
CJD International Support Alliance

There is no proven treatment for any of the prion disorders, particularly Variant CJD (vCJD). Four clinical trials of oral Quinacrine have taken place in the UK, France, Japan and the USA. The results of the French and the Japanese trials have been published. The UK trial has ended, with results yet to be published, and the USA trial is still recruiting patients..

Intraventricular infusion of Polyanion Pentosan Polysulphate (PPS) began in February 2003 in a small number of patients with prion disorders in various in Britain, Japan, France, North America and the European Union. A small number of papers have been published with more due this year.

PPS is well tolerated by this means of delivery and there have been no drug-related adverse side effects at the dosages administered.

Is PPS efficacious? We don't know yet although increased survivability has been demonstrated in certain strains.

In the UK, court injunctions restrict much of what can be reported on the patients treated. Injunctions have, however, been largely lifted in two cases:

- 1) Jonathan Simms (age 22)
- 2) Holly Mills (age 20)

Both patients were diagnosed with vCJD and both continue to receive intraventricular infusion of PPS around the clock. Jonathan started treatment in February 2003 and Holly began hers in November 2003.

Unlike sporadic CJD (sCJD), which has an average duration of about 6 months, vCJD has a duration of illness of around 14 months. Information in the public domain shows that all three vCJD patients in the UK undergoing PPS treatment have survived for more than 40 months- one reaching 64 months by Feb this year.

CJD Alliance UK held a successful Open TSE Discussion in Glasgow, Scotland, in December last year. Presentations were made by four key speakers and the minutes and presentations have been widely disseminated to groups including:

- UK Secretary of State for Health
- SEAC (Spongiform Encephalopathy Advisory Committee)
- Department of Health
- HPA (Health Protection Agency)

By means of such open dialogue, it is hoped that the relevant authorities will now acknowledge and act upon the information that came out of the discussion in Glasgow.

Our Electronic Mailing List

If you would like to be included in our electronic mailing list please email your address to:
s.solvyns@cjdsupport.org.au .

We are now in the position to send bulk emails of newsletters, invitations and information of interest to you via email. You have three choices.

- You can elect to only receive information, newsletters and invitations from us only via email (you will need to advise us of your email address).
- You can receive all information via email but still receive your newsletters and invitations by mail.
- Remain on our mailing list as before. If you do not contact us this is the way we will continue to inform and contact you.

Bulk emails will **not** make your name or email address available to be viewed by any other members of CJDSGN. If you are already receiving information from us via email it is not necessary to re-register for this service.



Bonded against CJD – United by Beads

These unique bracelets are part of an exclusive series of only 100 pieces created specially for the CJD Support Group Network, by artist Bianca Velder from South Australia. The glass beads are handmade and each bracelet is unique.

To purchase a bracelet and assist the CJDSGN to raise money for CJD Research please send your orders to Suzanne (s.solvyns@cjdsupport.org.au) or tel 1800 052466 – credit card facilities available.

\$25.00 each plus \$2.50 postage and handling

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Timetable 2007

February 2007

19th Feb – Committee Meeting

DVD production commenced

March 2007

10th March – NSW Support Group Meeting – Balmain Leagues Club

Guest speaker – Joe-anne Bendall, NSW Health Department.

Newsletter

May 2007

Meeting NT Health Department

NT Support Group Meeting

Committee Meeting

“Riding for Dad” Fundraising Event – Alice Springs

June 2007

Queensland Support Group Meetings

July 2007

1st July – “Hel’s Angels” Fundraising Event – Gold Coast

7 – 9th July – CJD Foundation USA Family Conference Washington DC

August 2007

Committee Meeting

Victorian Support Group Meetings

Tasmanian Support Group Meetings

September 2007

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October 2007

SA Support Group Meetings

WA Support Group Meetings

November 2007

9th November - Fundraising Gala dinner
Committee Meeting

