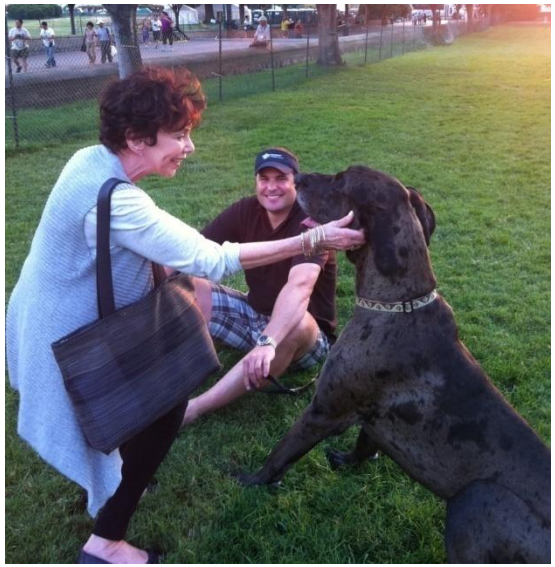
CJD International Support Alliance

CJD 2011 and the Ninth Annual CJD Foundation Family Conference

Washington DC
Sunday July 10th 2011



Who Are We?
A Brief History

Florence Kranitz

Co-Chairmen of the CJD International Support Alliance

In 2006 a group of CJD Support Foundation organization leaders from around the world decided to find a moment during a break in one of the hectic CJD Foundation Conference days to get together and talk to each other about our work.

I am still touched by the memory of that first meeting which took place on a hot Saturday afternoon in July in the busy lobby of this very Hotel. Our group gathered on chairs, the arms of chairs and the floor…. which just goes to show that it's the soul not the surroundings that drive matters of the heart.

There we were, representatives from organizations in Australia, England, Japan, Scotland, Northern Ireland and two from the U.S, all having been personally affected in some way by a prion disease. Despite a few language and accent barriers we discovered in each other the identical purpose and passion. Then and there we decided to organize an umbrella organization known today as the CJD International Support Alliance, and we pledged to work together on behalf those families touched by prion diseases around the world.

2006

The membership has grown and now, we have added organizations in Italy, France, and Israel and as of this past Thursday, we welcome Mexico.

As an Alliance we are concerned about food and blood safety in each country, clinical trials and promising research developments. We support each others causes by sharing information, issuing joint press releases and discussing issues of concern with our Board of Directors and our members. We try to act as a watchdog coalition speaking out on behalf of patients and families the world over, always remaining free from personal or political bias or prejudice. This group of closely bound member organizations, like you here at the conference, were total strangers to each other until we met and realized how we were all united by one mission that as survivors, our work is the most meaningful way we can honor the memories of those lost to CJD, those presently fighting and those, sadly, yet to come as well as those living with risk, the worried well.

Suzanne and I have been co chairmen almost since the group's inception and I can only tell you that working with her means having a partner I can totally depend on, when she gives her word she means it. No matter how full her CJDSGN schedule is she manages to carve out whatever it takes to do her share of the work. What would a night in Akron Ohio, late morning in Australia be without a flurry of email communications flying between us? Many times I have said g'nite only to find myself drawn back to our "email meeting" until midnight or later!

IN fact, Suzanne came to stay with me a few days before this conference and there we were both working at my kitchen table when suddenly I realized we were on the same side of the world in the same room, at the same table yet there we were emailing each other!!

The CJDISA meets via teleconference and once or twice a year face to face. Organizing the times for our teleconference continues to be a challenge, especially since some countries begin and end daylight savings time in different months and some don't change at all and even when we are all on standard time we are still between 7-13 hours apart. We are grateful to Suzanne for being the world clock time keeper.

Today you will hear a little about the work each of our Alliance partners and about some of the outreach Suzanne and I have carried out on behalf of the Alliance.

Now it is my great pleasure to introduce Suzanne, the National Co coordinator of the CJD Support Group Network, Australia and my Co-chair of the International CJD Support Alliance

What does the Alliance contribute to the global picture?

Suzanne Solvyns

Co-Chairmen of the CJD International Support Alliance

Working together to support all affected by Prion Disease around the World.

Since becoming an Alliance there has been a much more harmonious atmosphere between the research community and the patients, families and people at risk. For families to be able to reach out through the Alliance for information, or to better understand that answers cannot always be provided, seems to be dispelling some of the fear and anger and to know no matter where they are in the world they are not isolated . There is always someone who will answer their call for help within hours.

Often families will reach out to an overseas organisation for help but as an Alliance member we can refer them to the organisation that can offer them the most support. Sometimes a patient is in one country and the family in another so together we can provide the best assistance.

Another important benefit of networking is increased contact from families in countries where there is no support or formal organisation. They are often desperate and may reach out to several countries but we always try to help or refer them to where they can receive the best care‘.

We have developed a unity of support for all people affected by CJD and other prion diseases, Support networks/foundations have often been established in the past to offer support to specific groups. To support only familial CJD families or to only offer support for Iatrogenic cases or people at risk 'the worried well' but by joining forces and working together we are learning about the challenges that face each group. Deana from CJD Insight who offers support specifically to genetic family members has also helped with information and support to genetic families from other countries and I have spoken to and offered support to some of the worried well in USA.

Attending the CJD Foundation conference in 2005 coincided with the expansion of our Australian network from just providing support to people who were at risk after receiving human pituitary hormones to supporting all Australians affected by CJD and what we learned from CJD Foundation and others was invaluable . The Japanese began with support for patients and families affected by contaminated dura mater but have also expanded and I know that our French friends who offer support for families also affected by contaminated human pituitary hormones are also keen to expand and support other CJD families. We learn from each other so much, share resources and work as a team. We are of course very fortunate to have assistance from 21 experts and researchers our 'Friend and Advisors Group' and with their contribution and keenness to support all affected we really can make a difference.

Since our first meeting in Edinburgh in 2007 the Alliance has been asked to speak on behalf of those we represent in Madrid in 2008, Thessaloniki, Greece in 2009 Salzburg 2010 and in Montreal in 2011.

The conference in Montreal was excellent, extremely well organised and very well attended with almost 600 researchers, experts, scientists present. There were a lot of new faces and a lot of encouraging work presented. Our presentation was given a lunch time slot on Tuesday May 17, and so we were unsure if that would mean a large or small audience. To our delight lunch was a three course sit down meal so most if not all of the attendees were present. Our presentation followed a talk given by Honorable Greg Weadick, Minister of Advanced Education and Technology, Government of Alberta.

The audience were not as attentive as they should have been but I am happy to report that once Florence began to speak there was not a sound in the whole room and this continued during each segment of the personal stories with half if not more of the people in the room in tears. Florence prepared a wonderful talk which came from the heart and expressed some of the feeling that so many family members share with us and with you all during the work we do offering support. We then presented 4 personal accounts:
To view the presentation please visit either the home page of the CJDF or the CJDSGN
www.cjdfoundation.org www.cjdsupport.org.au

“Prion Diseases More than a Diagnosis"

Florence Kranitz

Suzanne Solvyns

Co-Chairmen of the CJD International Support Alliance

NeuroPrion 2011 Conference - Montreal Canada

Gary from USA shared the devastation for his family of the two year battle his wife Terry has had with CJD. Unfortunately Terry passed away on the night that we gave the presentation.

Maria-Gabriella from Italy shared a little of her tribute she has written and recorded after losing her father to CJD last year.

Amy from Australia gave an emotive account of dealing with her father's illness and then the horror of finding out that her father had a genetic mutation that she had also inherited.

Remy and Christel talked about the deaths of young people in France following treatment of human growth hormone and the stigma and fear for those who are at risk.

Our aim was to bring the faces of the people we represent to the meeting and touch the hearts of those present, thank them for their work and empathise just how important their work is to us all. If the reaction from the audience on the day and during the next couple days was anything to go by with both of us being thanked by people truly touched, I think we came close to what we aimed to achieve.

CJDISA
CJD International Support Alliance

CJD International Support Alliance

"Supporting those affected by Prion Disease the world over"

CJDISA
CJD International Support Alliance

Creutzfeldt-Jakob Disease Foundation, Inc.
U.S.A

CJD Support Network
Japan

CJD SUPPORT NETWORK
UK

CJD SUPPORT GROUP NETWORK
Australia

C.J.D.Israel

A.I.En.P ONLUS
Associazione Italiana Encefalopatie da Prioni
Italy

CJD Insight
U.S.A

"IN THIS TOGETHER"

Nous voulons juste grandir
MCJHCC
France

CJD Foundation Israel

Dear families,
My name is Alice Anane and I am the chairman of the CJD Foundation in Israel.
I apologize for not being able to attend this meeting personally.

Israel has dozens of patients who die each year from Prion Disease, and unfortunately more commonly it is the genetic form. Families are often shocked to discover that there is a family history and that they may also be a carrier of a genetic mutation for CJD. They are forced to learn about the consequences for themselves and their children. and often feel ashamed, living in denial they hide this information from other family members.

Despite the relatively large number of patients in Israel, we have a huge lack of awareness of the disease - even among medical doctors.

My father died of this disease when he was 49. Our family, existing of nine siblings, and my father's sister's family are all at risk of becoming sick with this disease. It is a very difficult living with this knowledge and we place very high expectations and hope in the research teams.
I founded the association in Israel with representatives of the families and researchers. Our goal is to raise funds in order to promote research and treatment, provide additional services such as support for families of patients as well as information and emotional support.

In 2009 our organization joined CJD International Support Alliance and we participate in the global enterprise and contribute to information-sharing at international level, consultation, support and advice. Alice

CJD Support Network Japan

A report from Japan

Our situation has not changed from the past years.

We were fortunate that the Touhoku earthquake and tsunami did not affect any of our members.
We continue to help through emails, phone calls and personal meetings, families who have suffered with CJD.

Let us strive and hope that the future will be better for us all.

Muneto

CJD Insight
USA

CJD Insight

"IN THIS TOGETHER"

CJD Insight

"IN THIS TOGETHER"

I apologize for having to leave early today but will share with you briefly a bit about CJD Insight

After losing my mom in 1998 to a disease I knew nothing about --- experiencing the helplessness, the frustration from lack of awareness, the pain of watching a vibrant women deteriorate, the knowledge of having lost 12 family members to familial CJD and knowing more will come, I was propelled to take action and work to prevent or ease the pain that other unsuspecting families would endure when blindside by this disease.

Coupling my personal experience, my nursing background and my work in information technology, I started the CJD Insight on-line web site/support group with a specific focus on familial CJD. I have had the blessing of working with many families across the world including many of you in the room today. We not only discuss how to care for a loved one or about the grief that comes with loss but also the very difficult topics of whether to get tested, knowing or not knowing genetic results, how to tell our children, concerns about access to care and insurance implications and how to live with the knowing or not knowing of a positive or negative result.

Very heavy stuff!

What I have discovered through my work is an emotional and spiritual bond that can never be severed and I have learned through all of you how to expand my own compassion and strengthen my own personal resolve. Thank you for being my source of energy, my inspiration, my 'balcony people' – it is for you and for my family that I do this work.

I would also like to thank the researchers for teaching me more than I ever wanted to know about science, research and genetics and for your undying dedication to this very rare group of diseases --- even before I knew this disease was in my family you were out there in the world working to find answers. Please never give up – we are here to encourage you and to put a face, a heart and a soul to the work you do.

Thanks also to the CJD Foundation, to all my professional colleagues of the CJD International Support Alliance and our Friends and Advisors. Unified we can make a difference and positively impact the families that we so passionately represent.

Remember – We are in this together and for that I am truly grateful.

God Bless!

CJD Support Group Network Australia

CJD Support Group Network Australia

David's Story

Why support was needed for recipients of human pituitary hormones in Australia

Thank you Florence

Good afternoon ladies and gentlemen

I have been asked to give a very brief account of how I became involved with CJD in Australia and how that has led to me being here with you at this conference.

Although I was not to know it at the time, my journey started nearly 40 years ago. At the age of 15, after my family doctor noticed that I was considerably shorter than my younger brother, and very short for my age, I was referred to the Endocrine Clinic at a large hospital in Sydney.

After a lengthy series of tests (some of which required stays in hospital while my friends were enjoying their school vacations), I was approved to receive injections of Human Growth Hormone. Fortunately for me, I responded very well to treatment, I appeared to have no side effects and I was able to complete my schooling, attend university and enjoy a very active sporting and social life.

As I entered my thirties everything was going well for me. I had met and married my lovely wife Lynette, we had the first two of what would become four beautiful daughters and my career was well on track. Then, in 1992, I received a letter from the National Department of Health which advised me to contact the Professor who had supervised my treatment all those years before. I had some idea of what it might be about because my mother had spoken to me after seeing stories in some of Australia's most prominent women's magazines which were reporting the deaths of Australian women who had been treated with Human Pituitary Hormones for infertility.

I made an appointment to return to the Endocrine Clinic, but before I attended my meeting, and so I was prepared to ask appropriate questions, I went to the library at the University of Sydney (where they have a well respected science and medical program) and I tried to research Creutzfeldt Jakob Disease and the possible side effects of hormone treatments. I could find nothing. It was as if this disease did not even exist.

I then prepared a whole page of questions that had come to mind and met with my Professor. He was friendly and he read a prepared script about four paragraphs long which advised me that some of the human hormone product that was used in the program had been contaminated, and confirmed that several Australians had died as a consequence of their treatment. As a result, I was at increased risk of developing CJD.

As it turned out, approximately 2100 Australians were informed that they were at increased risk of developing Creutzfeldt Jakob Disease as a result of treatment with human pituitary hormones either for short stature or infertility. Unfortunately for me, my questions to the Professor went largely unanswered. He either gave me very general responses or said that he did not know the answers to my questions. I left that meeting with little comfort and many issues that I needed to resolve. I visited my family doctor for advice, however, he had not even heard of CJD. In those days, not a lot was known and very little information was available.

The news that I was at increased risk of developing a rare and fatal disease for which there was no test and no cure changed my life. With my family's interest at heart, I put my career on hold, assumed a much larger role in the home and supported my wife as she went back to university to better prepare herself to support our family if the worst case eventuated and I was no longer around.
One difficult issue that I found myself confronting was that my parents, who thought that they were doing the right thing at the time when they gave consent for my treatment, were very worried and feeling terribly guilty that they had put me in this position.

In an attempt to find out more about CJD and to try and calm my mother, I attended some support group meetings in Sydney. I remember that first phone call in 1993 that I made to Suzanne who was our state coordinator at the time and finding out that she had been treated with pituitary hormones to assist with infertility and that she too was at increased risk of developing CJD. It was clear that there were many worried Australian recipients and their families and that very little was really known about this disease.

As a result of attending these support meetings, opportunities arose to participate in other state and national meetings and I joined Suzanne in being appointed as a recipient member of the National Human Pituitary Hormone Advisory Council, advising the Minister of Health at a national level.

When the Australian CJD Support Group restructured and expanded its role in 2004, from just supporting people who were at risk of CJD through hormone treatments, to providing support for Australians affected by all types of CJD, I was invited to join the management committee and I try to assist Suzanne, our tireless National Coordinator, where I can.

To help me keep abreast of the developments in the field of prion diseases (in particular, the areas of diagnostic testing, treatment trials and patient care) I have been fortunate to have been able to attend this outstanding family conference here in Washington for the past four years, for which I am truly grateful. As a result of meeting so many of you and hearing your stories, I have become very passionate about supporting families. Personally, I have learnt and grown so much as result of my visits here to Washington. I greatly appreciate the opportunity to attend and I especially appreciate the warm welcome I receive from Florence, your Foundation Board members and you the American families. I have no doubt that my experiences here have better prepared me to support our families back in Australia.

I would now like to introduce Suzanne, the National Coordinator of the CJD Support Group Network in Australia.

CJD Support Group Network Australia

Suzanne's Story

Expansion of the network to support CJD Families in Australia

Like David I have been involved in this Prion journey since 1992 when, being told of my risk. I felt like I was living a nightmare. At risk of a rare unheard of disease, to me anyway, with no treatment or cure and with very rapid onset!!!! This is the sort of stuff that horror movies are made of – not things that happen in real life. Why Me? How could the Australian government have endorsed the use of a product that was killing woman who had been on a fertility treatment, the same treatment I had received, a treatment that had also been given to 100's of young people to encourage growth. It seemed like madness but it was real.

I felt despair, I felt I had been delivered a death sentence, I feared for my life, for my family and believed that nobody could ever understand the despair I felt. I was wrong, so many of you in this room can relate to my story, to David's story. Those of you here who have lost a loved one, especially those of you who are dealing with a family history of this devastating group of diseases.

I had reached a door in my life, a door that I had to open to move on but I had no idea what lay beyond that door except for fear – I was scared and alone or so I thought. I moved on with a brave face but a heavy heart until the opportunity came to attend a meeting organised by the Australia health department to assist recipients of human pituitary hormones to establish a support network.

I was invited and went to see if I could help others, so I told myself, as it was hard to admit that I needed others, people who felt as I was feeling, people I could share my story with so that I did not feel so alone in dealing with this. I had plenty of support and love around me but no-one really understood!!!!

I met others, many very angry, others even more fearful that I was, many without good support systems and I saw how this news affected so many, parents of growth hormone recipients who felt guilty, sometimes their children blaming them, hidden truths, information coming via the media and often not accurate. Woman who had children on a program now with no idea if those children could be affected, suicides, marriage breakdown the list goes on.....

One thing became very clear if we linked arms and moved forward together as one voice providing mutual support it would help. A rocky road forward but made easier by taking that road together. If someone had told me then, almost 20 years ago, that I would stand here today relating my story and the story of why a support network was established in Australia I would not have believed it.

In 2004 I was contacted by a very irate young woman who had recently lost her father to familial CJD. She was angry that there was no official support for families. Her name was Mandy and some of you would have met her when she attended this conference in previous years. We had been very aware of the need of families and had been offering support where possible but Mandy's call emphasised the urgency of expansion.

The DoHA agreed in 2005 to amend our contract and provide appropriate funding so that support could be offered to all Australians affected by prion disease . We were also fortunately enough to attend the 2005 CJD Foundation 3rd Family Conference, to meet Florence Kranitz and many of you, family members. This was definitely the catalyst for a very fast expansion and the establishment of the CJDISA the following year bought unity and the sharing of resources that our families have certainly benefited from.

Since 2005 we have developed a data base for families, the number of families contacting us has steadily increased and this year, although Australia only has 25 – 35 cases a year we have already worked with 21 families, 17 of these have lost a loved one to highly possible or confirmed CJD.

We provide an information package for families, we network with health care professionals caring for a patient with suspected CJD and visit with patients and family members and assist in any way we can. In early 2008 we produced a DVD 'Understanding CJD' with assistance from Professor Colin Masters and Associate Professor Steven Collins and thank the CJDF who provided us with graphics. The DVD is a tool for our education program and since 2008 we have presented on 95 occasions to about 2500 health care professionals at in-services and conferences providing education and promoting awareness.

In 2010 with assistance from several dedicated and experienced health care professionals we produced a handbook 'Caring for a patient with suspected CJD' and have now distributed 1400 copies as part of an information package to hospitals all over Australia.

We have now established a data base of 1100 health care professionals who we network with when needed. We continue to advocate for timely access to health care for people at risk and family members. We have not eliminated delays but by maintaining pressure and being a voice for those at risk we are reducing discrimination.

Our funding to support families has been threatened since 2010 as funding comes from a trust account set up to support recipients of human pituitary hormones but we are hopeful that the criteria of the trust account will be altered so that our contract will once again reflect the actual work that we do. Attending this conference for the last 7 years and working with Florence has also meant for me the development of a great passion for our work with CJD families.

From a negative I believe we always have to try and find a positive and certainly Florence's talk last night was a wonderful example of that. I feel that this journey has given me a real purpose in life, a way to help others while helping myself. I have made so many new friends who are very dear to me, David, Florence and Jenny Cooke, journalist and author of Cannibals Cows and the CJD Catastophe who is here today, and of course so many of you experts in this field and family members alike and the families and hPH recipients in Australia. The legacy I want to leave is a well established and well known network that will continue to support all Australians affected by prion disease who will unfortunately come along in the future.
Thank you.

Now I will pass over to Roberto Borgis to tell you about the work of the organisation in Italy

A.I.En.P.
Italy


A I En P
ONLUS
Associazione Italiana Encefalopatie da Prioni

A.I.En.P.
Associazione Italiana Encefalopatie da Prioni

The Italian Association for Prion Encephalopaties – A.I.En.P.

CJD 2011 and the Ninth Annual CJD Foundation Family Conference

Washington D.C., July 8-11, 2011

Speaker: R. Borgis

Creutzfeldt-Jakob Disease Foundation, Inc.

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- Prion encephalopaties

  Epidemiological data

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Since **1993** a **National Register** for Prion diseases is active at Istituto Superiore di Sanità in Rome, Italy.

The system of surveillance on human encephalopathies was created in response to the spread of the epidemic of bovine spongiform encephalopathy that struck Britain at the beginning of the 90s.

The National Register ensures a single epidemiological analysis for a rare disease and keep monitoring the evolution of the sporadic and familial forms and any possible case of Bse.

In Italy the **majority of cases**, about **85 percent**, relate to the sporadic form, while the **familial forms** stand at **15 percent**.

Source: ISS web site

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		Numero di decessi in Italia (aggiornata al 30 giugno 2011)						
Anno	Segnalazioni	MCJ Sporadica	MCJ Iatrogena	MCJ Genetica	GSS°	FFI°°	MCJ Variante	Totale
1993	51	27	0	6	1	2	0	36
1994	62	33	0	6	0	1	0	40
1995	52	28	0	6	1	1	0	36
1996	77	51	0	7	0	1	0	59
1997	139	47	1	12	0	1	0	61
1998	143	64	2	9	0	1	0	76
1999	192	74	0	17	0	0	0	91
2000	177	60	0	23	4	3	0	90
2001	219	86	0	13	1	0	0	100
2002	201	77	0	11	3	0	0	91
2003	192	79	0	8	1	1	1	90
2004	176	78	0	20	0	0	0	98
2005	236	108	1	20	2	0	0	131
2006	238	96	1	31	5	2	0	135
2007	208	96	1	18	1	0	0	116
2008	217	92	0	12	0	0	0	104
2009	191	105	0	8	1	0	0	114
2010	216	93	1	10	6	0	0	110
2011	116	31	0	3	0	0	1	35

°GSS, sindrome di Gerstmann-Sträussler-Scheinker

°°FFI, Insonnia fatale familiare

Source: ISS web site

Updated June 30,2011

Creutzfeldt-Jakob Disease Foundation, Inc.

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A.I.En.P.
Associazione Italiana Encefalopatie da Prioni

- A brief history of A.I.En.P.

A.I.En.P.
Associazione Italiana Encefalopatie da Prioni

- **A.I.En.P. – the Italian Association on Prion Encephalopaties** is a **no-profit making association** formally established in **September 21, 2007 by 16 founding members** from North, Center and South of Italy; all are relatives (sons, daughters, husbands and wifes) of beloved ones stroke by prion encephalopaties, **in particular sporadic CJD and GSS;**

- The **headquarter** is in Rome;

- From 16 founding members A.I.En.P. reached **103 members.**

A.I.En.P.
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- The main activities in 2010 and the beginning of 2011

A.I.En.P.
Associazione Italiana Encefalopatie da Prioni

• **Contact with families**

During 2010 A.I.En.P. was brought into direct contact with families affected by prion diseases with an average of **3 phone calls per week**.

A.I.En.P. for each case and family gives both moral and concrete help, especially when "difficult" conditions are described deriving from uncorrect behaviour both by health professionals and hospitals.

• **Recognition as rare disease**

In March 2010 A.I.En.P. formally asked the Ministry of Health for the recogniction of prion diseases as rare diseases (which means economic relief for the families). The Ministry formally answered that our request is in stand by because the Ministry of Finance has to give a technical advice. It also underlined that only familial forms will be taken into consideration at this moment.

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- **Communication**

At the beginning of 2011 a renewed A.I.En.P. institutional web site is on line.

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• **Fund raising activities**

A.I.En.P. fund raising activities are active through:

- **members' annual fees**

-**donations by privates (individuals, corporate, institutions)**

- **5 per mille:** in 2010 A.I.En.P. received the amount of 2008 (9,671.43 eur – 233 choices) and received communication of the amount for 2009 (9,136,65 eur – 211 choices)

A.I.En.P.
Associazione Italiana Encefalopatie da Prioni

2nd Italian Family Day dedicated to prion diseases

Mario Negri Pharmacological Research Institute

Milan, December 4, 2010

A.I.En.P organised its second Italian meeting specifically dedicated to prion diseases. The meeting had scientific lectures in the morning and a round table in the afternoon in order to share experiences, ideas, best practice, etc.

The speakers were the most important scientific experts on prion diseases and come from Italy, USA, UK and Germany.

45 families attended the event.

Simultaneous translation was provided to all the attendees and speakers.

The participation was free of charge.

The video of the 2nd Italian Family Day dedicated to prion diseases can be downloaded on A.I.En.P. web site www.aienp.it

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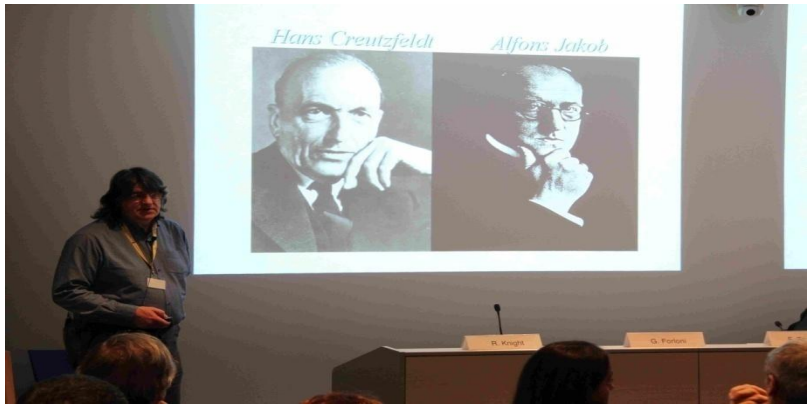

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A.I.En.P.
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In 2010 the Board of A.I.En.P. decided to give a **grant of 15,000 .00 Eur** to the Neuropathology Unit of the Neurological Institute Carlo Besta, Milan, coordinated by Fabrizio Tagliavini, **to support a researcher for the research activities on prion encephalopaties.**

The grant was focused on two innovative therapies on prion diseases coordinated by Dr. Fabrizio Moda. One work was published on *Nanoscale* and the other one received an honourable mention at he Student and post-doctoral fellow poster competiotion award at Prion 2011.

In 2011 the Board of A.I.En.P. decided for a new grant of 18,000.00 Euro to the Neuropathology Unit of the Neurological Institute Carlo Besta

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- The therapeutic value of writing

A.I.En.P.
Associazione Italiana Encefalopatie da Prioni

In 2009-2010 A.I.En.P had the privilege to witness the importance of writing in overcoming the pain of losing a loved one because of these terrible diseases.

2 of our members and a supporter wrote and published a book and a university thesis work.

Evalda Capirchio: ***The destiny of being rare – The voyage of Sergio Chillé*** - 2010
Memories of Sergio Chillé written by his wife (and founding member of A.I.En.P.).

Raffaele Pallavicino: *To you, who are part of me*, 2011
The story of Valeria, a special friend and a special friendship.

MariaGabriella Schirinzi: *CJD – One in a million*, 2008-2009. Thesis work for her University degree in Educational Sciences, Salento University

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MariaGabriella Schirinzi – CJD – One in a million.

MG in her University thesis work wrote the story of her father, Antonio Luigi Schirinzi, affected by sCJD and how the writing helped her in overcome the pain. This is her thought about the therapeutic value of writing to overcome the pain

Caring for a person is a real opportunity to increase our personal sense of life.

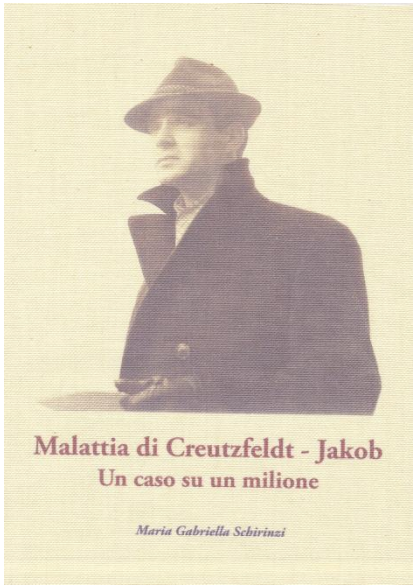
The disease can become a resource for both the sick person and for those around him, giving new meaning and value to life as a whole.

We can learn from our experience as caregivers that when life changes its color we have to look at it with different eyes, because there is a force within us that makes us absorb even the hardest shots and therefore change the way we see things and makes us go back to life.

To make sense of pain means to find elements of light and transform it into a place of growth. A wound that opens to light gradually can be healed and cured.
To overcome the pain we must open our hearts and minds.

To open up to others requires humility and courage, but then produces inner freedom, balance, opportunities to love and be loved.

*The **writing**, the autobiography of grief, the story telling is therapeutic and liberating and allows a relaboration of the trauma and mourning itself.*



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*Everything we **write** about us becomes our identity card. To decide to **write** your own story is a marker to start again our life's journey, to overcome the difficulties and the research for new destinations. **Writing** on ourselves is a hard work that gives you back energy and joie de vivre.*

***Writing** is like a drug; when you live there is always something to medicate. **Writing** conceived as art of remembering can be a therapy or even a cure. **Writing** gives consistence to memories.*

***Writing** is a self-healing method. It gives dignity, a history, a remembrance to those for whom it is said and written.*

***Writing** is a gesture of gratitude, of compensation, of symbolic restitution, to those who are not longer with us but left us a strong, beautiful and permanent memory in our lives.*

*Those whom we remember in our **written** memories become immortal.*

*To witness through **writing** our own experiences is a moment of collective participation and of renewal of memories of the loved ones who are no longer with us to the new generations.*

MariaGabriella Schirinzi

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France

MCJ-HCC
(Maladie de Creutzfeldt-Jakob

par Hormones de Croissance Contaminees)

JB Mathieu, President

CJDISA
CJD International Support Alliance

Nous voulions juste grandir
MCJHCC




To Jean-Philippe, Benedicte, Michael,

Nicolas and 116 other young victims of CJD…

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Nous voulions juste grandir
MCJHCC

MILESTONES - 1

1958: First method of extraction of of human Growth Hormone (hGH) from pituitary glands by Dr Raben in Boston

1973: First hGH treatments in France by France-Hypophyse, a non-profit organization of pediatric experts

1976: First warning on the risks of CJD transmission by hGH (by Dr Dickinson in UK)

1979: Genentech announces the availability of a biosynthetic GH molecule

November 1984: Death of Joe Rodriguez, 19, in San Francisco, first identified victim of CJD by hGH.

April 1985: Prohibition of the use of hGH, following the death of 3 patients.

December 1991: Death of Illyasil Benziane, first identified victim of CJD by hGH in France

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MILESTONES - 2

1993-1997: Seven physicians and pharmacists from France-Hypophyse are indicted for “involuntary manslaughter” and “deceit on

1996: “Pour Benedicte”, by Francine Delbrel

2002: “Le dossier noir des hormones de croissance”, by F. Delbrel and JB Mathieu

May 2008: First trial of the “Growth Hormones scandal”

October 2008: Death of Pr Job, President of France Hypophyse

2009: Tribunal Correctionnel of Paris clears all indicted “Not guilty”

Xmas 2009: Death of Fernando Meireles, last identified victim of CJD by hGH in France

2010: Appeal – All surviving indicted are cleared

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THE WAYS FORWARD

1- SUPPORT TO THE "WELL BUT WORRIED"

982 persons have been treated between 1983 and 1985 with suspicious batches of hGH. 120 have died, 862 have not developed CJD. The support to the survivors is a natural mission of our organization

2- SUPPORT TO OTHER VICTIMS OF CJD

We want to use the experience gained in handling CJD victims (in-house hospitalization, daily care, procurement of medical materials, administrative issues, etc…) and we are recruiting volunteers to help the families confronted to CJD. We are seeking help and guidance from the ISA Alliance for this task.

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MCJHCC

CJD Support Network
UK

UK
THE CJD SUPPORT NETWORK

Richard Knight

CJD SUPPORT NETWORK

Speaking on behalf of Gillian Turner

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CJD SUPPORT NETWORK

Speaking on behalf of Gillian Turner

Co-Chair of their Management Committee

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CJD SUPPORT NETWORK

Speaking on behalf of Gillian Turner

Member of their Management Committee

A Clinical Neurologist dealing with CJD

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CJD SUPPORT NETWORK

Established 1994

By relatives of people with CJD

For all forms of CJD

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PAST & PRESENT FUNDING

DONATIONS

UK Department of Health

NCJDSU

CJDISA
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FUTURE FUNDING ?

DONATIONS

UK Department of Health

NCJDSU

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CJD SUPPORT NETWORK

A PATIENT SUPPORT GROUP

Help and Support for:

People with CJD

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Help and Support for:

People with CJD

Their Carers

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Help and Support for:

People with CJD
Their Carers
Those at ‘Higher Risk’

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Help and Support for:

People with CJD

Their Carers

Those at ‘Higher Risk’

Concerned Professionals

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CJD SUPPORT NETWORK

Practical and Emotional Support

Information
Promoting good care for those with CJD
Financial support (caring grants)
Linking families with similar experiences
Running a national helpline

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CJD SUPPORT NETWORK

Providing information about CJD

Through:

Information sheets

Leaflets

Newsletters

Website

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CJD SUPPORT NETWORK

INFORMATION BOOKLET
MARCH 2007

CJD and prion disease

An introduction and explanation

CJD Helpline 01630 673973 www.cjdsupport.net

CJD SUPPORT NETWORK

INFORMATION SHEET 1 JANUARY 2008

Sporadic CJD

Sporadic CJD (sCJD) is one of the four different forms of Creutzfeldt-Jakob disease, which belongs to a group of rare, and always fatal, brain disorders called the prion diseases. These occur in both humans and animals, and include BSE. sCJD is also referred to as classical CJD.

CJD is caused by the accumulation in the brain of an abnormal form of a protein called a "prion protein". PrP can exist in two forms – normal (PrP^{C}) and abnormal (PrP^{Sc}). We all have normal PrP^{C} in our brain. The abnormal prion is different because it is folded in a different way and has a different shape to the normal. Abnormal prion protein can cause normal prion protein to change shape and become abnormal. This leads to a chain reaction which, in turn leads to damage of brain cells.

Introduction to CJD

CJD was first described in the 1920s by two German neurologists (Creutzfeldt and Jakob). It causes a progressive loss of mental abilities and is accompanied by neurological symptoms such as unsteadiness and clumsiness.

The disease affects about one person in a million per year, giving rise to 50 or so new cases a year in the UK. Of these, 85 per cent are sporadic, having no known cause, with the remainder comprising genetic, iatrogenic and variant (see information sheets 2, 3 and 4). sCJD is most common in the 45-75 age group, with the peak age of onset being 60-65.

At present, CJD can only be diagnosed for certain by post- mortem examination of the brain. Under a microscope, brain tissue from someone who had CJD has a characteristic spongy appearance, caused by numerous tiny holes where cells have died. For this reason, CJD, BSE and other prion diseases are sometimes called spongiform encephalopathies.

Microphotograph of spongiform change in brain tissue taken from a person with CJD

Variant CJD

Variant CJD (vCJD) was initially named New variant CJD. It is a new form of Creutzfeldt-Jakob disease, which belongs to a group of rare, and always fatal, brain disorders called the prion diseases. vCJD is generally believed to be caused by exposure to BSE, a prion disease found in cattle.

CJD is caused by the accumulation in the brain of an abnormal form of a protein called "prion protein". PrP can exist in – normal (PrP^{C}) and abnormal (PrP^{Sc}) forms. We all have normal PrP^{C} in our brain. The abnormal prion is different because it is folded in an unusual way and has a different shape to the normal. Abnormal prion protein can cause normal prion protein to change shape and become abnormal. This leads to a chain reaction which, in turn leads to damage to brain cells

A new type of prion disease

In 1995, two cases of CJD were found among teenagers in the UK. This was extremely unusual, and alarming, for only four cases of CJD (one in Britain) had ever been reported in this age group previously. By 1996 the number had increased to ten, and it was evident that a new type of prion disease, called variant (v) CJD, had arrived in Britain. The occurrence of an epidemic of BSE among UK cattle from 1986 was thought to be no coincidence. vCJD was soon linked to exposure to BSE prior to the 1989 ban on specified offal (brain and spinal cord) from cattle in the human food supply. Exposure may have continued via spinal cord in mechanically recovered meat until 1995.

Microphotograph of spongiform change in brain tissue taken from a person with CJD

The number of deaths per year due to vCJD in the UK increased from 1995 up to 2000 (when there were 28 deaths) and currently (2008) remains in decline with five deaths in 2007. By January 2008 there had been a total of 163 (definite and probable) deaths from of vCJD in the UK (with three patients still alive). Eight countries have reported between one and three cases, the Republic of Ireland has reported 4 and France 23 cases. Some of these non-UK cases are thought to have arisen during stays in the UK, while others are thought to have arisen within the relevant country.

CJD SUPPORT NETWORK

Genetic CJD

Genetic CJD (previously called familial CJD and sometimes referred to as inherited CJD) is an inherited form of Creutzfeldt-Jakob disease, which belongs to a group of rare, and always fatal, brain disorders called the prion diseases. These occur in both humans and animals, and include BSE and scrapie in animals.

Introduction to genetic CJD

CJD is caused by the accumulation in the brain of an abnormal form of a protein called a "prion protein". PrP can exist in two forms – normal (PrP^{C}) and abnormal (PrP^{Sc}). We all have normal PrP^{C} in our brain. The abnormal prion is different because it is folded in a different way and has a different shape to the normal. Abnormal prion protein can cause normal prion protein to change shape and become abnormal. This leads to a chain reaction which, in turn leads to damage of brain cells

Microphotograph of spongiform change in brain tissue taken from a person with CJD

Genetic CJD accounts for around 15 per cent of all cases of CJD. There are fewer than five new cases occurring in the UK each year. Like the other forms of CJD, genetic CJD is characterised by dementia (mental decline with symptoms such as memory loss) and neurological problems such as unsteadiness. The brain of someone with genetic CJD will also show the spongiform change which is the hallmark of all forms of CJD - the brain tissue has a spongy appearance when viewed under a microscope.

Genetic CJD is caused by a genetic mistake where a mutation in the PrP gene seems to make the conversion into the abnormal form more likely. Several different mutations have now been identified. There are also two other, even rarer, inherited brain diseases which resemble genetic CJD. These are Gerstmann Straussler Scheinker disease (GSS) and fatal familial insomnia (FFI). Like genetic CJD, they are associated with mutations of the PrP gene. The distinction between these different forms of disease (GSS, FFI and genetic CJD) is partly historical and currently many experts tend to class these diseases together under 'genetic prion diseases'.

Inheriting a risk of CJD

We all inherit two copies of the PrP gene – one from our mother, and one from our father. Genetic CJD, GSS and FFI are all inherited in an autosomal dominant fashion. That is, you need to possess just one mutated copy of the PrP gene to develop the disease. A person carrying the mutated gene has a 50 per cent chance of passing it on to each child. Since CJD does not usually strike until later in life, people carrying the gene may not realise that they may have passed it on to their children, although they may well be aware of a problem with neurological disease within the family.

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Iatrogenic CJD

Iatrogenic CJD (iCJD) is a form of Creutzfeldt-Jakob disease, which belongs to a group of rare, and always fatal, brain disorders called the prion diseases. This form of CJD arises from contamination with tissue from an infected person, usually as the result of a medical procedure.

CJD is caused by the accumulation in the brain of an abnormal form of a protein called a "prion protein". PrP can exist in two forms – normal (PrP^{C}) and abnormal (PrP^{Sc}). We all have normal PrP^{C} in our brain. The abnormal prion is different because it is folded in a different way and has a different shape to the normal. Abnormal prion protein can cause normal prion protein to change shape and become abnormal. This leads to a chain reaction which, in turn leads to damage of brain cell.

Our first awareness of prion diseases

The first indication that human prion diseases might be transmissible through infected tissue came with the discovery of a strange disease called kuru among the Fore people of Papua New Guinea in the 1950s. Kuru mainly affected women and children, and began with unsteadiness of gait, shakiness and lack of co-ordination. Behavioural changes followed, although dementia was unusual (making it different from sporadic CJD). Eventually the patient would become unable to move and death would follow, usually within a year of onset of symptoms. The brains of these patients showed severe damage to the cerebellum, the part of the brain which controls movement. There were also spongiform changes (characteristic of prion disease) where the brain tissue has a spongy appearance when viewed under the microscope. A further feature was the appearance of small deposits called plaques within the brain tissue, distinguishing kuru from CJD, where plaques only occur in a minority of cases.

Microphotograph of spongiform change in brain tissue taken from a person with CJD

Kuru was eventually linked to the funeral practices of the Fore people, in which it was common for the women and children to handle the body of their dead relatives, including the brain. Whether they practiced cannibalism, as has been widely reported, and actually ate the brain is not known. But if just one member of the tribe had sporadic CJD, any woman or child handling brain tissue could have been contaminated by it merely through scrapes or scratches on their body. Since the victims of kuru went on to be given these funeral rites, the disease perpetuated itself.

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Variant CJD and blood transfusions

Why am I at risk of developing vCJD?

You have a higher risk of developing vCJD than the general public because you received blood from a donor who either went on to develop vCJD themselves or you gave blood to another person who later developed vCJD. We know that it is possible for vCJD to be transmitted through blood.

What is the likelihood of developing vCJD? What is the risk?

Unfortunately, we don't yet know the precise risk. Most people in the UK are at some risk of developing vCJD because they have eaten beef or beef products contaminated with BSE in the past (see below, vCJD and BSE). We know that your risk is rather higher because you received this particular donor's blood. For this reason, the Health Protection Agency (HPA) has informed you that you are 'at risk for public health purposes' and that you need to take certain precautions to protect other people from this increased risk.

It is not clear whether everyone infected with vCJD will develop symptoms. It is possible that the disease will remain 'asymptomatic' (not have any symptoms in some people and will not affect their health.

1. vCJD and BSE

Both BSE (bovine spongiform encephalopathy, sometimes referred to in the press as 'mad cow disease') and vCJD are members of a group of diseases called TSEs (transmissible spongiform encephalopathies). TSEs are caused by a highly infectious agent called a 'prion', and are also known as prion diseases.

Before the mid-1990s there were few controls on cattle feed or on the content of mechanically recovered meat used in pies, hamburgers, sausages and 'ready meals'. Some cattle were fed with tissue from animals infected with a prion disease and they subsequently developed BSE. Tissue from the brains and spinal cords of these BSE-infected cattle were included in mechanically recovered meat, which was consumed by members of the general public. Some of these people went on to develop vCJD, which is believed to be the human form of BSE.

How soon will I know?

Sadly, we're not sure. Very few people have been infected with vCJD through a blood transfusion. We know that this has occurred in four cases. (March 2007) In three of these the person receiving the blood went on to develop vCJD. The length of time between the blood being received and the person showing definite signs of vCJD (the incubation period) appears to have been in excess of six years.

where can I get more information and support?

The information that you are 'at risk of CJD for public health purposes' has probably come as a shock. You may feel isolated, bewildered and unsure of where to obtain further information and support.

There are a number of agencies that can provide you with more detailed information. In particular, we would encourage you to contact the CJD Support Network (helpline: 01630 673973; website: www.cjdsupport.net email: info@cjdsupport.net). The Network offers practical and emotional support for those diagnosed with any form of CJD, those considered at risk of developing CJD, their friends and families.

Contacts

CJD Support Network
PO Box 346
Market Drayton
Shropshire
TF9 4WN

Telephone 01630 673973 (helpline)
Telephone 01630 673993 (admin)
Website: www.cjdsupport.net
Email: info@cjdsupport.net

National Prion Clinic
PO Box 98
National Hospital for Neurology
and Neurosurgery
Queen Square
London
WC1N 3BG

Telephone 020 7405 0755 (direct line)
Fax: 020 7061 9889
Website: www.nationalprionclinic.org
Email: help.prion@uclh.org

Human BSE Foundation
Helpline: 0191 3890 04157
Website: www.hbsef.org.uk

National CJD Surveillance Unit
Western General Hospital
Crewe Road
Edinburgh
EH4 2XU

Telephone: 0131 537 2128 (clinical office)
Telephone: 0131 537 1980 (pathology)
Fax: 0131 343 1404
Website: www.cjd.ed.ac.uk

Health Protection Agency (HPA)
CJD Section
61 Colindale Avenue
Colindale
London
NW9 5EQ

Telephone: 020 8327 7418/6075/6074
Fax: 020 8200 7868 (FAO CJD Section)
Website: hpa.org.uk/infections/topics_az/cjd/menu.htm
Email: kate.soldan@hpa.org.uk

CJD SUPPORT NETWORK

INFORMATION SHEET 7 OCTOBER 2001

Aggressive behaviour in CJD

If you are caring for someone with Creutzfeldt-Jakob disease (CJD) you may find they behave in an aggressive way which is uncharacteristic of their usual personality. You may be surprised, upset and wonder how best to cope.

If a person with CJD reacts with aggression, it is probably because they feel frightened, humiliated or frustrated by what is happening to them. Since CJD is such a rare condition it may be that the person affected picks up on the confusion of their carers, who may not realise what is happening.

Because CJD affects the brain, judgment and self-control may be eroded. The person affected may forget all about their normal habits of good behaviour and manner learned in childhood. They might completely over-react to any criticism or comment you make – however mild – of their altered behaviour. Obviously it's upsetting for carers to be attacked verbally, or even physically, in this way. But it's important for you to remember that the aggression is not directed to you personally. In fact, the person with CJD will probably forget the incident much faster than you do.

Triggers for aggression

There is often a trigger for aggressive behaviour. If you are caring for someone with CJD, you may find it worthwhile to keep a diary of events. This is a useful way of learning what upsets the person affected which, in turn, makes it easier for you to plan a strategy to prevent it happening. Some pointers to watch out for are:

- Feelings of frustration or humiliation because the person can no longer cope with everyday life. Since people with variant CJD are often in the younger age-groups, there may be a very marked contrast with their usual lifestyle.
- Concerns about loss of privacy and independence as others take over intimate functions like washing, dressing and going to the toilet.

CJD SUPPORT NETWORK

INFORMATION SHEET 5 JUNE 2001

Swallowing problems

The brain damage caused by prion disease sometimes causes swallowing problems which are distressing for patient and carers alike These problems may also lead to malnutrition. If eating and/or swallowing become difficult, it is important to ask your GP for a referral to a speech and language therapist for advice.

The process of swallowing

Swallowing gets food from the mouth through the throat (pharynx) and down the oesophagus into the stomach. The throat divides into two tubes at its base – the one at the front is the windpipe, leading to the lungs, and the one at the back is the oesophagus. It is vital that food doesn't get diverted into the windpipe, otherwise breathing may be obstructed. To stop this happening, there is a reflex which causes the windpipe to close while food passes down the oesophagus. Should any food accidentally enter the windpipe, coughing – another reflex – should remove it.

Swallowing is a three-stage process:

- In the oral preparation stage, the lips, tongue, cheeks and teeth work together, breaking food into a soft ball that can readily be swallowed. The tongue forms a cupped shape around liquids and holds them ready to swallow.
- During the pharyngeal stage, the tongue squeezes food or liquid to the back of the mouth, which triggers the swallow reflex, temporarily closing the windpipe. Muscles in the wall of the throat help food and liquid pass into the oesophagus by a set of wave-like movements called peristalsis. Once the food is safely past the opening of the windpipe, it re-opens.
- During the final – or oesophagal – phase, food and drink complete their journey down the oesophagus to the stomach, aided by further peristaltic waves.

Brain damage may affect any of these stages. For instance, muscle movement in the throat could become slow or lacking in co-ordination. The swallow or cough reflex may be affected or the

CJD SUPPORT NETWORK

INFORMATION SHEET 6 JUNE 2001

The autopsy in patients with suspected CJD

Answers to some commonly asked questions

Why are post mortem examinations requested in patients with CJD?

Although a diagnosis of probable or possible CJD can be made during the clinical illness in individual patients, the only way to confirm the diagnosis is by performing a post mortem examination of the brain. This information is important in establishing the incidence of CJD in the UK. Performing a post mortem examination also allows the opportunity to study these devastating diseases – how the brain is damaged, the changes occurring in other parts of the body and scientific studies on the nature of the agent causing these diseases.

Is it necessary to remove the entire brain during the post mortem examination?

A diagnosis of CJD can sometimes be confirmed by examining only a small portion of brain tissue -- this can also be done in life by performing a brain biopsy. However, this can give a number of potentially misleading results. It is internationally accepted that the best way of confirming a diagnosis of CJD is to retain the brain at the time of post mortem for full examination in the laboratory. During the post mortem, the brain is carefully removed (using an approach from the back of the head) and most of the tissue is fixed in formaldehyde. Some of the tissue may be frozen in order to perform biochemical studies to identify the abnormal prion protein in the brain.

How can I give consent for a post mortem examination?

A new consent form for post mortem examinations was established in 2000 on the basis of guidelines from the Royal College of Pathologists. Using this form, relatives are asked for permission to perform an autopsy on a patient with suspected CJD and then asked for specific permission to retain the brain for diagnosis. Additional permission is also requested to use the brain tissue for research and teaching purposes so that more can be found out about these diseases. Relatives will be asked if they would like to have the brain tissue returned for subsequent burial or cremation.

CJD SUPPORT NETWORK

Helpline +44 (0)1630 673973

www.cjdsupport.net

Newsletter

Issue 18

March 2009

CREUTZFELDT-JAKOB DISEASE SUPPORT NETWORK NEWSLETTER

Is dental treatment a risk factor for variant CJD?

A dental study update

Pauline Watt and Fiona Ord, Research Dental Hygienists, NCJDSU, Edinburgh

This dental study began in April 2008 and has been funded by the Department of Health to assess whether dental treatment is a potential source of transmission of vCJD. To date, there is no evidence of transmission of vCJD through dental treatments.

In June 2008, letters were sent to families to gather names and addresses of the dentists where their relatives were registered. With the information received from families, it has been possible to contact many dentists to obtain as much information as possible about the dental treatments carried out.

This information is being collated along with the dental treatments of approximately 650 controls for comparison.

Dental practices are required to keep patient records for a number of years, however the dental treatments of some of the relatives have been destroyed. For those that have been destroyed the NHS Practitioner Services in Scotland and England/Wales are able to provide a record of treatments received, if claimed under the NHS. This information is stored as far back as 1998 and has been useful in building a history of treatments where the original records are not available. Much time and effort goes into locating dental practices and records as unlike medical records, dental records do not follow patients if they move from one practice to another. The collection of dental treatments is still ongoing and work has begun on inputting this information into a statistical database.

Informative articles have been published in dental journals to raise awareness of the study and poster presentations will be submitted for consideration at dental conferences in 2009/2010.

In the coming year, the information will be gathered and entered into a database and the results will be published. These will be distributed to committees e.g. SEAC and to organisations developing policies relating to public health issues.

NCJDSU would like to express their gratitude to all the families who have provided information. If anyone has not heard from NCJDSU regarding the study and would like to provide information on their relatives dentist, please contact Pauline or Fiona on 0131 537 2251/2345.

Published by CJD Support Network, PO Box 346, Market Drayton, TF9 4WN · Telephone 01630 673993
Email info@cjdsupport.net · Website www.cjdsupport.net · Charity number 1097173

CJD SUPPORT NETWORK

Website

Text, Audio, Video, Podcast

www.cjdsupport.net

April 2010-March 2011: 58,800 Unique Visitors

40 unique visitors used website to ask questions via email

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CJD SUPPORT NETWORK

Promoting good quality care for those with CJD

Providing training, workshops, conferences

Encouraging adoption of good practice guidelines

Campaigning through regular contact with ministers and senior government officials

Promoting research into CJD

CJDISA
CJD International Support Alliance

CJD SUPPORT NETWORK

Family Support Day

Information including invited talks

Q & A Sessions

CJDISA
CJD International Support Alliance

Running a National Helpline

CJD Support Network helpline

01630 673973

THE NATIONAL HELP LINE

“I NEEDED TO SPEAK TO SOMEONE AT 9 pm”

“I NEEDED TO SPEAK TO SOMEONE ON SUNDAY”

CJDISA
CJD International Support Alliance

Phone Help Line
April 2010-March 2011 : 527 Calls

Affected Families	**299**
Blood Recipients 'At Risk'	**31**

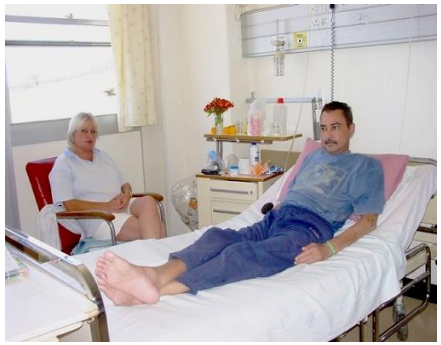
Worried Well

- **Health Sector**
- **Media**
- **Social Services**
- **Other**

CJDISA
CJD International Support Alliance

RELATIONSHIPS

CJD
SUPPORT
NETWORK



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RELATIONSHIPS

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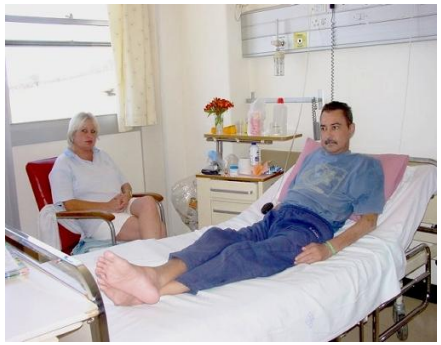

Gillian Turner

CJD INCIDENTS PANEL
CJD RESOURCES OVERSIGHT COMMITTEE

CJDISA
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RELATIONSHIPS

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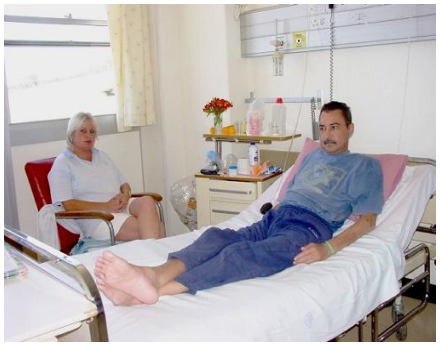

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Helpline +44 (0)1630 673973 www.cjdsupport.net

Newsletter Issue 18 March 2009

CREUTZFELDT-JAKOB DISEASE SUPPORT NETWORK NEWSLETTER

Is dental treatment a risk factor for variant CJD?

A dental study update

Pauline Watt and Fiona Ord, Research Dental Hygienists, NCJDSU, Edinburgh

This dental study began in April 2008 and has been funded by the Department of Health to assess whether dental treatment is a potential source of transmission of vCJD. To date, there is no evidence of transmission of vCJD through dental treatments.

In June 2008, letters were sent to families to gather names and addresses of the dentists where their relatives were registered. With the information received from families, it has been possible to contact many dentists to obtain as much information as possible about the dental treatments carried out.

This information is being collated along with the dental treatments of approximately 650 controls for comparison.

Dental practices are required to keep patient records for a number of years, however the dental treatments of some of the relatives have been destroyed. For those that have been destroyed the NHS Practitioner Services in Scotland and England/Wales are able to provide a record of treatments received, if claimed under the NHS. This information is stored as far back as 1998 and has been useful in building a history of treatments where the original records are not available. Much time and effort goes into locating dental practices and records as unlike medical records, dental records do not follow patients if they move from one practice to another. The collection of dental treatments is still ongoing and work has begun on inputting this information into a statistical database.

Informative articles have been published in dental journals to raise awareness of the study and poster presentations will be submitted for consideration at dental conferences in 2009/2010.

In the coming year, the information will be gathered and entered into a database and the results will be published. These will be distributed to committees e.g. SEAC and to organisations developing policies relating to public health issues.

NCJDSU would like to express their gratitude to all the families who have provided information. If anyone has not heard from NCJDSU regarding the study and would like to provide information on their relatives dentist, please contact Pauline or Fiona on 0131 537 2251/2345.

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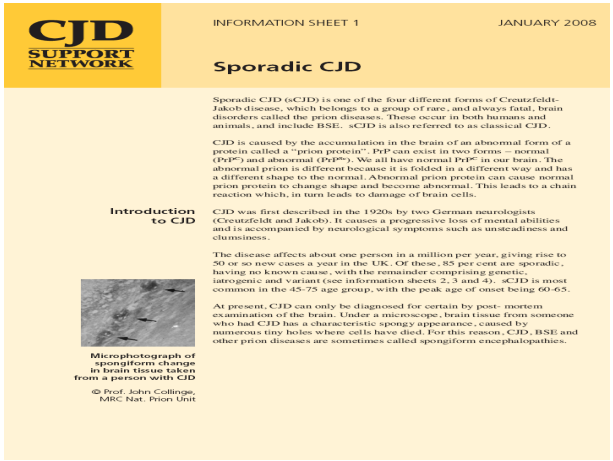

CJD SUPPORT NETWORK

INFORMATION SHEET 1 JANUARY 2008

Sporadic CJD

Sporadic CJD (sCJD) is one of the four different forms of Creutzfeldt-Jakob disease, which belongs to a group of rare, and always fatal, brain disorders called the prion diseases. These occur in both humans and animals, and include BSE. sCJD is also referred to as classical CJD.

CJD is caused by the accumulation in the brain of an abnormal form of a protein called a "prion protein". PrP can exist in two forms – normal (PrP^C) and abnormal (PrP^{Sc}). We all have normal PrP^C in our brain. The abnormal prion is different because it is folded in a different way and has a different shape to the normal. Abnormal prion protein can cause normal prion protein to change shape and become abnormal. This leads to a chain reaction which, in turn leads to damage of brain cells.

Introduction to CJD

CJD was first described in the 1920s by two German neurologists (Creutzfeldt and Jakob). It causes a progressive loss of mental abilities and is accompanied by neurological symptoms such as unsteadiness and clumsiness.

The disease affects about one person in a million per year, giving rise to 50 or so new cases a year in the UK. Of these, 85 per cent are sporadic, having no known cause, with the remainder comprising genetic, iatrogenic and variant (see information sheets 2, 3 and 4). sCJD is most common in the 45–75 age group, with the peak age of onset being 60–65.

At present, CJD can only be diagnosed for certain by post- mortem examination of the brain. Under a microscope, brain tissue from someone who had CJD has a characteristic spongy appearance, caused by numerous tiny holes where cells have died. For this reason, CJD, BSE and other prion diseases are sometimes called spongiform encephalopathies.

Microphotograph of spongiform change in brain tissue taken from a person with CJD

© Prof. John Collinge, MRC Nat. Prion Unit

RELATIONSHIPS

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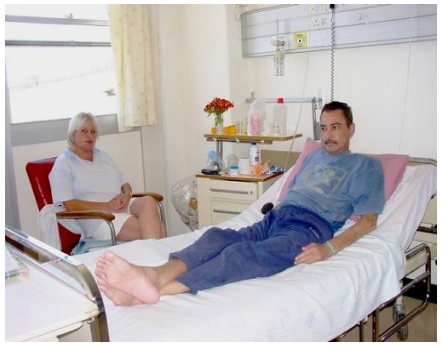

COMPLAINTS

WISHES

QUESTIONS

WORRIES

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RELATIONSHIPS

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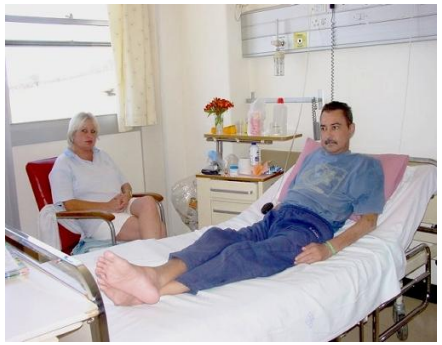

SOME COMPLAINTS

SOME WISHES

SOME WORRIES

MAY BE UNDERSTOOD BETTER BY NON-PROFESSIONALS

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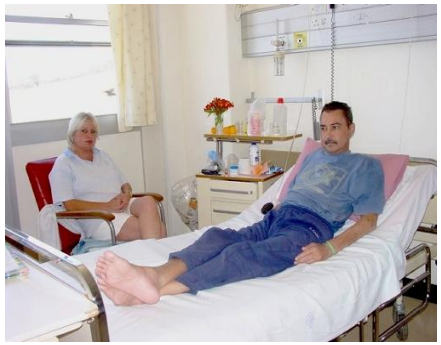

NOT *ALL* COMPLAINTS ARE JUSTIFIED

NOT *ALL* WISHES ARE REASONABLE

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CJD SUPPORT NETWORK

MANAGEMENT COMMITTEE MEMBERS WHO HAVE TURNED THEIR PERSONAL GRIEF INTO POSITIVE ENERGY TO HELP OTHERS

CJDISA
CJD International Support Alliance

CJD Foundation Mexico

CJD Surveillance in Mexico

TECNOLÓGICO DE MONTERREY
Escuela de Medicina
Campus Guadalajara

UNIVERSIDAD DE GUADALAJARA
PIENSA Y TRABAJA

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CJD Surveillance in Mexico

Population: c. 105 million

120 million

Health coverage:

- IMSS (workers) ISSSTE (gov. employees)
- SSA (non workers) Private

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CJD Surveillance in Mexico

HOT SPOTS

- Prevalence
  - State/national review (death certificates, path. archives)*
- Patient referral
  - Letters to neurologists
  - Web page
- Diagnosis
  - Level 2+ laboratory*
- Support
  - Official status*
  - Grants

*INDRE (National Institute for Epidemiological Referral)

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CJD Surveillance in Mexico

Working Group on Prions

- Composed of Federal and Public Health Institutions, Universities.
  - Ministry of Health
  - COFEPRIS (Federal Council for Health Risks Prevention)
  - SAGARPA (Ministry of Agriculture)
  - INDRE (National Institute for Epidemiological Referral)
  - IMSS (Mexican Institute of Social Security)
  - National Institutes of Health (National Institute of Neurology and Neurosurgery)
  - IPN (National Polytechnic Institute)
  - TEC Guadalajara/UDG (Me)

Cofepris
Comisión Federal para la Protección contra Riesgos Sanitarios

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CJD Surveillance in Mexico

GOALS

- Established as a Working Group for the creation of a Mexican statement on Canadian Beef
  - Meeting with the Canadian Working Group
- To create a Risk Assessment for Mexican Beef
  - Mainly by SAGARPA/COFEPRIS
- To support the Surveillance
  - Laboratory
  - Law, legal status
- To create a National Epidemiological Assessment
  - Epidemiological referral

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CJD Surveillance in Mexico

ACHIEVEMENTS

- Mexican statement on Canadian Beef
  - Cancel the ban on Canadian beef
- Revalidation of OIE status
  - Safety of Mexican beef

PERSPECTIVES

- To support the Surveillance
  - Laboratory
  - Law, legal status
- To create a National Epidemiological Assessment
  - Epidemiological referral

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PATIENT REFERRAL

- Peers (direct communication)
  - USA: CJD Foundation
    NPDPSC
  - UK: NCJDSU
  - Germany: NRZ
- Contact by the internet
  - Web page: www.fundacioncjd.com
- Referrals from local neurologists
- Referrals, abroad
  - Spain, Costa Rica, Panama, Colombia

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OVERVIEW ON CJD ASSESMENT

- Doctors with limited experience
- Referral: 4-5 months
- Most cases have been extensively studied (MRI, EEG, …)

- Misconceptions:
  - + 14-3-3 = CJD
  - MRI analysis limited (pulvinar)

  - A CJD patient must be isolated
  - Brain biopsy must be done

  - Genetic counseling overlooked

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Relatives

- Looking for information concerning CJD
  - What are Prions, CJD
  - Inheritance
  - Infectivity
  - Diagnosis (is it really CJD?)
  - Treatment
  - Prevention (Vaccination)

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CJD Surveillance in Mexico

RESOLUTIONS

- Promote educational outreach to doctors with limited experience
- Provide lectures on diagnostic process and testing options

- Provide general information that is understandable to laypeople or non-scientific professionals

- Foundation to become incorporated, non profit organization
- Recruit volunteers and support system

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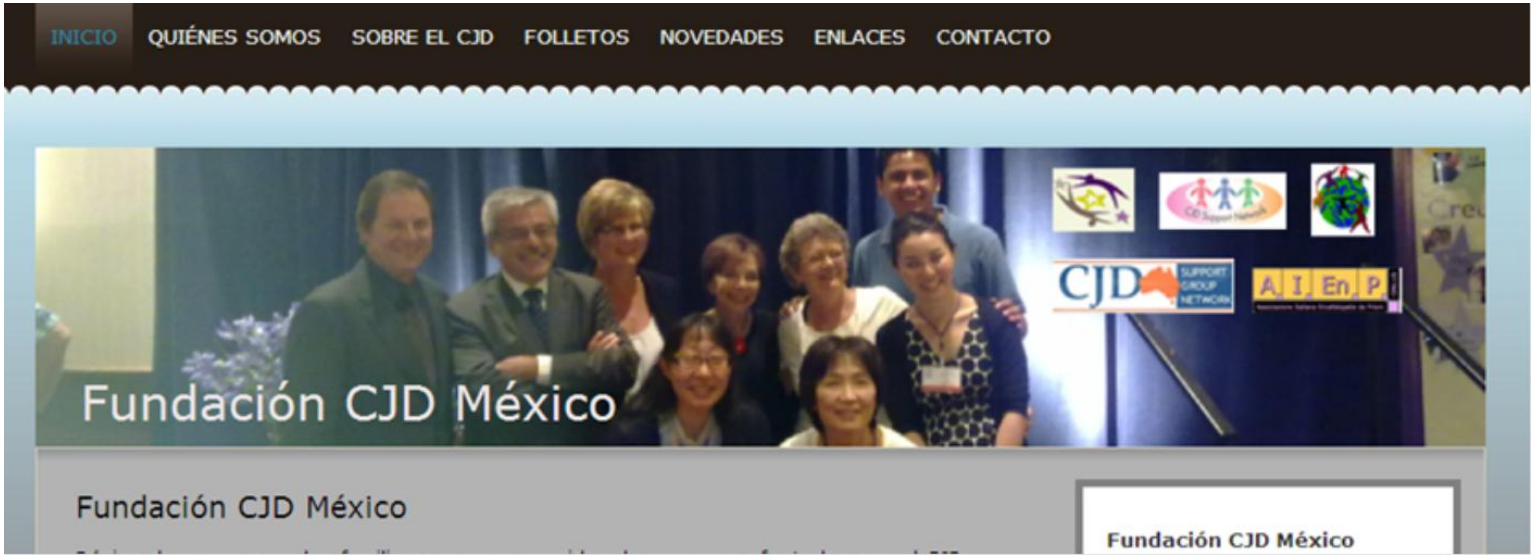
CJD Surveillance in Mexico



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Florence Kranitz, CJD Foundation
Suzanne Solvyns, CJD Support Group Network
International Support Alliance
Physicians and caregivers who refer cases

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Questions & Answers