



Creutzfeldt-Jakob Disease
Foundation, Inc.

The CJD Foundation Newsletter

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President's Message

Dear Friends,

I apologize for the length of this newsletter but I have a lot to share with you.

Please note that Thursday, November 12 is International CJD Day. Clearly those of us who have either lost a loved one to a prion disease or are presently caring for a family member or friend diagnosed with a suspected prion disease, do not need a special day to remember, however, we hope that you will honor your loved one by helping to educate others about this tragic family of diseases. Roseann Bray, one of our CJD Foundation family members has written her thoughts about this poignant day, you can read it on our website www.cjdfoundation.org. We also want to thank Roseann for speaking about CJD at a continuing education seminar for funeral professionals in Pennsylvania.

On September 23-25, Marisa and I attended *Prion 2009* held in Thessaloniki, Greece. Each year NeuroPrion, the European Network of Excellence, sponsors a scientific meeting consisting of 2 1/2 days of presentations by preeminent prion disease scientists, new researchers and, in some cases, students who have worked on promising new scientific projects. There were more than 500 prion disease scientists in attendance at this year's meeting, most of them specifically focusing on prion disease research.

While in Greece we welcomed two new CJD support organizations, one from Israel which focuses on supporting families affected by familial CJD and one from France supporting families who have lost a young family member to CJD acquired through contaminated human growth hormone injections. This growing global community presently consists of organizations from Australia, France, Israel, Italy, Japan, the United Kingdom, and, of course, the United States. It was very moving to have scientists from Israel and France seek us out to introduce us to representatives of their countries' CJD Foundations who had traveled to Greece specifically for this purpose.

Suzanne Solvyms, my CJD International Support Alliance Co-Chairman and I were honored to be invited to address the assembly. Below is a copy of my report written for the NeuroPrion newsletter:

Building a Global Prion Disease Support Community

The CJD International Support Alliance (CJDISA), made up of not-for-profit prion disease support organizations around the world, was privileged to address the audience of distinguished prion disease researchers in attendance at *Prion2009* in Thessaloniki Greece on September 25, 2009.

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The Alliance presentation offered insight into their growing role pointing out that this growth is due, in many respects, to the support and encouragement of the prion disease research community. Since its inception three years ago, the CJDISA has developed a strong network which now has the ability to refer families affected by prion disease to medical professionals in almost every country, provide information and connect families to support mechanisms all over the world. These connections provide the building blocks for a global prion disease support community.

During their presentation, the CJDISA expressed its deep gratitude to the scientific community and to Alliance Bio-Secure and Dr. Valerie De Broglie for sponsoring our attendance at this important meeting. They ended with a brief video showing patients and families affected by a prion disease. This video brought the human face of this tragic category of diseases directly to those who work daily to find treatments and, hopefully soon, a cure.

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We came away knowing that there is a lot of very hopeful research being conducted all over the world. You can access the NeuroPrion newsletter soon at www.neuroprion.org/en/np-newsletters.html. The presentation abstracts are currently posted on www.neuroprion.org

Following the meeting in Greece, Marisa came back to Akron to “hold down the fort” while I traveled to Milan with Suzanne and David Ralston (Australia), to attend the first European Union (EU) Family Conference. Raffaella Robello and Roberto Borgis, officers of the Italian CJD Foundation (Associazione Italiana Encefalopatie da Prioni) planned and carried out this extremely successful meeting, inspired by our CJD Foundation Family Conferences, which brought families together from all over Europe. The language barrier was successfully managed through simultaneous interpreters. Seeing family members who had felt so alone and isolated all together in that room was extremely moving. The day consisted of excellent presentations by ten leading prion disease experts followed by a question and answer period. The participation from the audience highlighted just how hungry they were for information.

I trust that the feeling everyone went away with was one of hope and connection and willingness to attend again whenever and wherever the next EU meeting is held. Suzanne and I were part of the planning committee and we both felt very privileged to have had a role in this important day. Bravo to A.I.En.P and to Raffaella and Roberto for such a fine conference!

Upon returning to the CJD Foundation office, I heard the exciting news that Dr. Pierluigi Gambetti, Director of the National Prion Disease Pathology Surveillance Center and our Medical Director, had received an award from the Annals of Neurology for his recent discovery of a new strain of prion disease. The new disease is named “protein sensitive prionopathy or PSPr,” and there is some suggestion that it may be the long sought sporadic form of GSS. This prestigious award was presented to Dr. Gambetti at a ceremony in Baltimore, MD on October 13.

Continuing with good news, as most of you know the CJD Foundation is available through our HelpLine and website email address to families from all over the world. We offer referrals, practical help and general information. Unfortunately, however, at times we are unable to adequately communicate due to the language barrier, and at these times we turn to friends of the Foundation for help. Recently Dr. Wen-Quan Zou, Associate Director of the National Prion Disease Pathology Surveillance Center who is a premiere prion disease researcher as well as a clinical neurologist, offered to help us and has accepted the role of CJD Foundation International Consultant. Dr. Zou is currently in the process of translating our website into Chinese (there is not even a word for CJD in the Chinese language) and he has offered to make himself available to families contacting us from China or other Asian countries. In the few weeks since he began, Dr. Zou has already been able to offer advice to families in Viet Nam and The Philippines. We are very grateful for this much needed help.

We also want to thank Leticia Harrison, a family member, for volunteering to translate our newsletters into Spanish. The translations of our May and August newsletters are posted on our website.

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...President's Message continued

The information packets handed out at *Prion 2009* contained information about our two new grant opportunities and we have subsequently received 33 Letters of Intent! Three are applications for the GSS Grant funded by the Yobs family in honor of their husband and father, Patrick Yobs and the second is underwritten by the families of Albert Tennant and Paul Oberdorf for human prion disease research. If you are interested in helping underwrite a research grant, please contact us for further information.

We are already working on *CJD 2010 and the Eighth Annual CJD Foundation Family Conference*. You should have received your "Save the Date" email last Friday. Please plan to join us in Washington, July 16-19. Monday, July 19 is designated as Advocacy Day on Capitol Hill.

We are happy to announce two new CJD Foundation Board of Directors members, Veronica Oberdorf and Paul Paternoster. Both Veronica and Paul have worked very hard on our behalf and we look forward to their valuable input as Board members.

This Saturday, November 14, the Snively family is holding *Footwork for CJD* in honor of their wife and mother, Marsha Snively. Proceeds will be donated to the CJD Foundation. The event, a walk, is being held in Fairmont, West Virginia. The Snively's are hopeful that this event will provide comfort to everyone participating in memory of a loved one lost to CJD. For more information please contact Nikki Bland at bland_nikki@yahoo.com.

In closing, I send my warmest regards to each one of you and hope that as we move into the holiday season you are able to spend time with family and friends and that you find meaningful ways to incorporate memories of past holiday celebrations into the present, thus finding peace and healing.

Most sincerely,

Florence



Questionnaire Update

Thanks to our families, the CJD Foundation's questionnaire efforts continue to be very successful. The questionnaire was revised in September 2008 at which time we started mailing them with our condolence packets. Since the revision and mailing began, we have received 403 completed questionnaires! All of the information provided, including that dating back to 2000, have been entered into our database and are being analyzed on an ongoing basis by our epidemiologist consultant, Steven Korzeniewski. Steven began compiling his report using those questionnaires received from September 2008 through March 2009 and presented the results at CJD 2009 and the *CJD Foundation Family Conference*. Steven's presentation is available on NeuroPrion's website: http://www.neuropriion.org/en/video_090712_03.html. He will now begin work on the earlier data and will issue an updated report in the months to come. With Steven's expertise, we are planning to submit this body of work for publication in the next few months

We are asking family members who returned their questionnaire prior to receiving final autopsy results to please contact us so we can complete your information in our database. If possible, we would like you to attach a copy of the autopsy report to your questionnaire. If you are unable to send a hard copy, please call or email us with the information - it is important to include these findings in our data analysis. Reports can be faxed to 330.668.2474, emailed to help@cjd.foundation.org, or mailed to The CJD Foundation, P.O. Box 5312, Akron, OH 44334. Please feel free to email or call us with any questions or if you need help obtaining the results. As always, this information is confidential and randomized in all reports, no names are ever used.

We look for possible trends or similarities in patient backgrounds and for any other information which might prove helpful to our researchers. The Questionnaire not only provides a unique overview but it also offers each family, who is willing to share their story, a safe and meaningful way to do so. At the present time, we are the only repository of anecdotal CJD patient information in the United States. If you have not yet completed a questionnaire and are willing to do so, please contact us at 1-800-659-1991 or help@cjd.foundation.org.

HelpLine Statistics

The CJD Foundation's largest ongoing project is our toll-free HelpLine (1-800-659-1991). The HelpLine is available to families coping with a loved one's suspected CJD diagnosis who are in need of support and information, to medical professionals, funeral directors and embalmers, the general public, and the press and the media. Below you will find HelpLine statistics for January 1, 2009 – October 31, 2009.

The CJD Foundation is not a reporting agency and families are not required to report their loved one's illness or death to us. These statistics are not intended to be scientific in nature, but instead to help validate the work we do on a daily basis.

2009	NEW CASES RE-PORTED	U.S. DEATHS REPORTED	HELPLINE (FAMILY) CALLS	HELPLINE (FAMILY) EMAILS	GENERAL INQUIRY CONTACTS	MEDICAL PROF CONTACTS	UNIQUE WEBSITE VISITORS
January	26	26	84	76	8	12	4,788
February	15	15	67	70	3	12	4,993
March	29	22	108	82	12	9	5,462
April	23	20	67	88	8	8	4,994
May	29	20	80	50	4	7	5,554
June	12	12	64	60	-	5	4,717
July	27	20	62	33	2	7	4,086
August	20	21	67	42	5	2	4,550
September	25	13	54	26	4	2	4,411
October	16	10	62	45	1	11	4,147
TOTALS =	222	179	715	572	47	75	47,702

Note 1: Not all new cases and deaths reported are confirmed by autopsy.

Note 2: "Contacts" include emails and phone calls.

CJD 2009 and the CJD Foundation Family Conference Speaker Presentations

The CJD Foundation wishes to express extreme gratitude to NeuroPrion, the European Network of Excellence, for sending two videographers from their headquarters in Paris once again this year to film *CJD 2009 and the Seventh Annual CJD Foundation Family Conference* and for allowing us to link to their website to view the presentations. The support and friendship of NeuroPrion has been, and continues to be, invaluable. We are grateful to them for their belief in us and in our CJD International Support Alliance partners.

Please click this link the following link to view the presentations:

<http://www.neuroprion.org/en/patients-events-09-CJD-Day.html>

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House Approves Food Safety Bill

Congress took a major step toward improving the safety of the American food supply in July, when the House of Representatives approved bipartisan legislation (H.R. 2749) to overhaul the food safety system.

"If approved by Congress and signed into law, this bill would go far toward reducing the toll of food-borne illness in the United States," said Christopher Waldrop, Director of CFA's Food Safety Institute.

Testifying before the House Agriculture Committee in July on behalf of a dozen food safety organizations, Carol Tucker Foreman, Distinguished Fellow of CFA's Food Policy Institute, said the bill "includes the key elements most frequently noted by experts as essential to an effective food safety system."

In an historic first, the bill specifically directs the Food and Drug Administration (FDA) to take steps to prevent foodborne illness, and it assigns resources to begin the job.

It would, for example, require all food companies to develop systems of preventative controls, require the FDA to develop minimum safety standards for production of fresh fruits and vegetables, allow the FDA access to relevant food safety records and increase the frequency of inspections at higher risk facilities, and impose civil penalties.

"The bill would make food safety a primary focus of the FDA, not an afterthought as it has been for so long," Waldrop said.

"The time has come for Congress to act responsibly, consider the interests of those who consume food as well as those who produce and process it, and pass the Food Safety and Enhancement Act promptly," Tucker Foreman added.

House passage came on a 283-142 vote. The focus now moves to the Senate, which is expected to consider food safety legislation in the fall.

Meanwhile, CFA participated in a meeting in May with chairs of the newly created White House Food Safety Working Group, which is exploring ideas on ways to attack the problem of foodborne illness.

In a letter to President Obama, CFA and other consumer and public health groups expressed appreciation that the administration is giving the issue such prominent attention. "No previous President has acted so early in his Administration or adopted such an open process to address food safety concerns, and for that we are grateful," the groups wrote.

The groups detailed actions in four areas that they urged the Administration to take:

1. working with Congress to secure passage this year of legislation that gives the FDA the authority and tools it needs to assure the safety of the foods it regulates;
2. developing legislation to reform and modernize the federal meat and poultry inspection system;
3. working with Congress to address the need for basic structural reform of the food safety regulatory agencies; and

adding several citizen representatives to the White House Food Safety Council.

"We are encouraged by the focus of the Administration and Congress on food safety. By the end of this year, we hope the President will have signed strong food safety legislation into law," Waldrop said.

This article was originally published in the Consumer Federation of America's June-August 2009 newsletter, CFA News

Podcast

The CJD Foundation's first podcast is available on www.cjdfoundation.org. Thank you to Dr. Brian Appleby and Deana Simpson!

Our second podcast is scheduled to be recorded on Wednesday, November 11th. Dr. Pierluigi Gambetti will be interviewed by Dr. Brian Appleby. Please check our website to listen to the new podcast!

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