

## **CJD/Prion Diseases Questions and Answers**

Note – Due to time limitations during the 4/2/14 Grand Rounds, the following questions could not be addressed by the speakers, Michael Fischer, MD, DSHS has kindly provided the following responses.

***Question: I saw a National Institutes of Health documentary which indicated that even when exposed to fire, the prion was not destroyed. Also, it was indicated that it continued to exist even when buried. Is that still accurate?***

Answer: In the context of the topic presented the phrase “exposure to fire” is vague. Putting a hamburger on the grill and cooking it to well-done would not be sufficient to destroy the prion (the temperature is too low, the pressure applied is at atmosphere, and the duration of exposure is too short). Now, incinerating contaminated materials is sufficient in destroying/deactivating the abnormal prion protein. The temperature, pressure, and duration of exposure are sufficient for destruction/deactivation of the prion protein.

Incineration is the only known way to be certain that the prion protein is completely deactivated (destroyed). The disease causing form of the prion protein is considered resistant to radiation, heat, and chemical treatments alone. The temperature and length of exposure required to destroy the agent is higher and longer than the standard decontamination and sterilization measures used for surgical equipment. A combination of chemical and heat treatment laid out by the WHO guidance has been determined to be acceptable for the destruction of the pathogenic form of the prion protein. The protocol and guidance recommended by the WHO and CDC can be found at: [http://www.cdc.gov/ncidod/dvrd/cjd/ga\\_cjd\\_infection\\_control.htm](http://www.cdc.gov/ncidod/dvrd/cjd/ga_cjd_infection_control.htm)

In general proteins are stable and have long half-lives under normal conditions, this is the same for the abnormal prion protein and it will exist for an extended period of time buried or not buried. Refer to the CDC website regarding infection control and the message to funeral home directors. “CJD is not transmissible from person-to-person by normal contact or through environmental contamination. For example, it is not spread by airborne droplets as are tuberculosis (TB) and influenza or by blood or sexual contact as are hepatitis and human immunodeficiency virus (HIV). CJD transmission can occur during invasive medical procedures involving the central nervous system due to exposure to contaminated brain tissue. This accounts for less than one percent of all CJD cases. ([http://www.cdc.gov/ncidod/dvrd/cjd/funeral\\_directors.htm](http://www.cdc.gov/ncidod/dvrd/cjd/funeral_directors.htm))

***Question: Can you please explain familial CJD and plain genetic CJD?***

Answer: In general, the terms Genetic CJD( or prion disease), Familial CJD, and Inheritable CJD are synonymous terms and are used interchangeably. These terms are referring to a disease caused by the same protein by the same mechanism of action but the big difference is that these diseases are associated with a mutation in the gene that codes for the prion protein (the PRNP gene). There are several mutations in the prion protein that are passed on from parents to offspring in an autosomal dominant pattern, thus the use of the term inheritable. Familial CJD (fCJD), Gerstmann-Straussler- Scheinker Disease (GSS), and Fatal Familial Insomnia (FFI) are the genetic/familial/inheritable prion diseases. These group of disease typically have an earlier onset (patient become symptomatic at an earlier age) and the duration of illness is longer. Predominant features of the illness vary with the type of mutation.

**Question: For Patient 24, mentioned in the Grand Rounds presentation, what was determined as the point of infection?**

Answer: This patient was determined to have sporadic CJD (sCJD). There is no event or environmental factor linked or associated with occurrence of sporadic CJD. It is believed that this is a spontaneous occurrence and not acquired, so to the best of our knowledge there is no point of infection to be determined (1 death per million population per year which is a relatively stable rate throughout the world).

**Question: Can you give us a description of the types of current CJD research that is being conducted?**

Answer: I think Dr. Ances' review of drug trials are good example of research done. In addition, there are studies being done to better understand the normal functioning of the human prion protein, some of this is done with zebra fish. Other current studies include the RT-QuIC test which is now being reported following the results of the 14-3-3 and tau protein tests if consistent with CJD. The RT-QuIC test stands for Real Time- Quaking Induced Conversion [of the normal prion protein to the abnormal form]. As this test becomes more accepted and fine-tuned, more studies to delineate time of infectivity and incubation periods will be looked at, as well as familial CJD and how early can we test for progression of disease (can we pick up the occurrence of the disease process prior to onset of symptoms?). I am unaware of any drug trials going on at the present time. I would suggest checking out the [cjd.foundation.edu](http://cjd.foundation.edu) website, look for the page on the CJD family conference and click on presentations. Many of the presentations are on current research.

**Question: Can the disease be transmitted through body fluids.**

Answer: The abnormal prion protein has been identified in bodily fluids.

**Question: I understand that CJD is a required reportable disease in Texas. How many states nationwide have required reporting of CJD? Are there plans to require reporting in all states?**

Answer: I am not sure of the precise number of state which have made CJD notifiable, but most of them have. It has been proposed in the past, I am not aware of any active requests for making CJD a nationally notifiable disease.

**Question: Are there precautions a family member of a prion infected person should take?**

Answer: No. As per the WHO guidance, "Normal social and clinical contact, and non-invasive clinical investigations (e.g. x-ray imaging procedures) with TSE patients do not present a risk to healthcare workers, relatives, or the community. There is no reason to defer, deny, or in any way discourage the admission of a person with a TSE into any healthcare setting. Based on current knowledge, isolation of patients is not necessary; they can be nursed in the open ward using Standard Precautions." (The document can be viewed at:

[http://www.who.int/csr/resources/publications/bse/WHO\\_CDS\\_CSRAPH\\_2000\\_3/en/](http://www.who.int/csr/resources/publications/bse/WHO_CDS_CSRAPH_2000_3/en/)

**Question: Is the incidence of CJD similar throughout the world? Are there any variance based on racial or cultural groups?**

Answer: One death per million populations per year is a relatively stable rate that is seen throughout the world. There are exceptions and for the most part this is be attributed to a familial/genetic CJD being higher in that population rather than an increase in sporadic CJD or acquired CJD.

***Question: What are the parallel studies or research in scrapie (ovine)? Any zoonotic / human cases from scrapie?***

Answer: Yes there are lots of studies comparing human prion disease with scrapie, both the similarities and the differences. The list is very extensive. The occurrence of Scrapie can be traced back to the 1700s, there are no known cases of transmission to humans even though there has been hundreds of years of exposure.