

## **Creutzfeldt-Jakob Disease**

### ***An informative review with suggested guidelines for care and handling of the dead prior to cremation***

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The following is an overview of the emerging issue of handling the dead human body with Creutzfeldt-Jakob Disease. The purpose is to educate the reader to what the disease is and is not and to better prepare the funeral service professional cremationist to service clientele while protecting the public and employees of the crematory. Creutzfeldt-Jakob Disease (CJD) is a relatively new disease having been first identified by researchers naming the syndrome after themselves around 1920. The last three years has seen a dramatic increase in the awareness of the disease. However, many researchers agree that worldwide that there has not been a true increase in the number of new cases. The attention has in many instances confirmed existing cases of CJD. These cases may have gone undetected or misdiagnosed without the acute worldwide attention the disease has gained.

CJD is by all accounts a rare, 100% fatal brain disorder that causes a progressive dementia and numerous neuromuscular disorders. In the United States the Centers for Disease Control and Prevention in Atlanta (CDC) continue to identify the cases of CJD in the range of one case per 800,000 to 1,000,000 deaths. CJD statistically develops in individuals after the fourth decade of life, with the bulk of cases being identified in the range at 45 to 75 years of age. CJD has been shown to lay dormant in the body for decades showing no symptoms during this long incubation period. For reasons not fully understood but potentially linked to aging and the decline of the immune system, the disease will begin to manifest itself with tell-tale signs of forgetfulness, anxiety, depression, and varying behavior changes.

The cause of CJD was undetermined until recent developments identified the true makeup of the causative agent. Dr. Stanley Prusiner, professor of neurology and biochemistry at the University of California, San Francisco won the 1997 Nobel Prize for medicine for his work on prions which has opened a new and alarming world of infectious disease. Prions are now added to the list of agents that cause disease along with bacteria, viruses, fungi, and parasites. A prion is said to stand for 'proteinaceous infectious particle' and it is the structural makeup of the prion that makes it act differently than any other known infectious agent. Prions are proteins that are found in both normal and abnormal forms but appear not to contain nucleic acids. Nucleic acids are found in all living things regardless of the level of complexity and are the foundation of DNA and RNA structures of all living things. The abnormal structural form appears to be responsible for transmitting biological mechanisms which may be responsible for a multitude of dementia related illnesses. Furthermore, the abnormal configuration of the prion is what makes it so ominous and difficult to eradicate. The prion is believed to have an effect on normal protein molecules converting them into lethal forms by causing the once normal protein to change shape and function. Prions are resistant to a wide variety of chemical and physical disinfectants and can withstand wide temperature ranges and still be potentially disease causing. The modes of infection of the CJD prion in humans is not well understood at present and there is no test to detect a patient's immunologic status to the presence of the CJD prion. Researchers are

developing tests for the future where a small amount of lymph tissue may be recovered from a tonsil or like tissue for biochemical assay, but these tests are purely experimental at present.

It is known that the disease can be introduced in laboratory animals by percutaneous inoculation of infective material such as brain tissue or cerebral spinal fluid from an infected person. Transmission in numerous cases has been associated with the use of contaminated surgical instruments which have become contaminated during brain surgery, corneal transplants, and dura mater grafts. There is documentation of transmission of the disease following inoculation of contaminated pituitary derived growth hormone as well. Most importantly, person-to-person transmission via skin contact or via environmental contamination has not been shown.

At present, there are several classifications of the disease based upon how researchers believe the disease is transmitted:

#### **INHERITED CREUTZFELDT-JAKOB DISEASE**

It is believed that as many as 20% of all known CJD events are inherited. These cases are known as familial cases. Statistical information pertaining to confirmed cases of CJD have identified increased familial presence in communities of individuals around the world including the United States. Ancestor related disease in those born in Czechoslovakia, Chile, and Libyan-born Jews are also well documented.

#### **SPORADIC CREUTZFELDT-JAKOB DISEASE**

Those individuals who have developed CJD but have no known exposure to the infectious agent and where there is no evidence of the disease in the family in past generations are referred to as sporadic. At present, most cases of CJD are considered sporadic.

#### **CREUTZFELDT-JAKOB DISEASE BY EXPOSURE**

While the agent responsible for CJD is believed to be the prion, it is not considered contagious in the conventional meaning of the term. No proof exists that demonstrates a direct link of the development of the disease in confirmed cases of individuals who have worked in allied health fields. At present, the only proven way of contracting the disease is from an infected person who has been through medical procedures which have used tainted surgical instruments or infected tissue for transplant and therefore the infection was direct. Further, research has found no higher incidence rates of CJD in family members or spouses living with or caring for a patient with CJD. This does not preclude taking strict precautions when handling potentially infectious tissue of a CJD patient. Strict universal precautions must be followed at all times when in contact with the body of a suspected or known CJD case.

#### **NEW VARIANT CREUTZFELDT-JAKOB DISEASE**

Studies released in October of 1997 have confirmed a new variant form of CJD, known as nvCJD, and it is believed to be the same strain of prion causing the much written about bovine spongiform encephalopathy (BSE), or 'mad cow disease' found in the United Kingdom in cattle. Based upon past studies performed in primates, lower laboratory animals and biochemical studies, researchers suggest that nvCJD is the human counterpart of BSE. At present no cases of nvCJD have been identified in the United States.

Regardless of the type of CJD diagnosed, CJD is not recognizable by external observation. At present the only definitive methods of diagnosis remain antemortem biopsy of cerebral tissue which is costly and potentially harmful to the patient and therefore not performed routinely or the post-mortem examination of the brain. CJD, unlike other diseases, does not produce fever, wasting of the body, or changes in the skin texture or color. With all that is known about CJD, what remains unclear is the precise method(s) of potential exposure which may result in disease development at some point in the future. The following are suggested minimum guidelines for handling the bodies of deceased CJD patients. Upon review, the reader will notice that at the forefront of exposure reduction is the use of strict universal precautions, which has been recommended repeatedly for use in varying degrees during any contact with all dead human remains.

### **SUGGESTED GUIDELINES**

- \* Funeral homes should be advised that a body with CJD must be prepared for cremation prior to delivery and acceptance at the crematory as the cremation container will not be opened for any reason upon delivery
- \* Since embalming has no effect on reducing the pathogenicity of the prion, embalming should not be mandatory for acceptance for cremation
- \* All containers/caskets used for cremation must be leak-proof
- \* All bodies should be placed in a cadaver bag or heavy duty disaster pouch prior to being placed into the cremation container/casket
- \* Strict use of full body, universal precautions must be followed at all times the body is present; including eye, nose and mouth protection; double gloving, hair and shoe covers, full length fluid-proof gowns
- \* Percutaneous exposure to blood, brain tissue, or cerebral spinal fluid of an infected person should be immediately followed by irrigation of the wound with sodium hypochlorite (household bleach)
- \* Any skin contact with possibly infectious materials should be followed by washing with 2 N sodium hydroxide
- \* Any fluid leakage should be immediately decontaminated using copious amounts of 2 N sodium hydroxide. Following treatment with 2 N sodium hydroxide, the area should be mopped with disinfectant soap and water and rinsed thoroughly.

As with all emerging issues, the previous discourse is based upon the best scientific evidence available at the time of the writing. The author and the Association will continue to monitor all aspects of the issues and updates will be made available in a timely fashion.