Prion Diseases

AN OVERVIEW FOR FUNERAL SERVICE PROFESSIONALS

BY MEGHAN LEWIS, KATIE GLISIC, DANIELLE JORDAN AND DR. BRIAN APPLEBY

Mortuary professionals have encountered many potentially fatal and infectious diseases and have successfully found ways to both serve families and protect the health of the community. Prion disease is no exception. Prion diseases, also referred to as transmissible spongiform encephalopathies (TSE), occur in humans and animals and primarily affect the central nervous system. The disease occurs when normal proteins spontaneously change to abnormal, disease-causing proteins.

There are three prion disease classifications: sporadic, genetic and acquired. The majority are of sporadic etiology (85%) and include sporadic Creutzfeldt-Jakob Disease, sporadic familial insomnia and variably protease-sensitive prionopathy. Genetic prion diseases account for 5% to 10% of cases and comprise familial Creutzfeldt-Jakob Disease, fatal familial insomnia and Gerstmann-Sträussler-Scheinker.

Finally, the remaining cases are acquired through either ritualistic cannibalism (kuru), ingestion of prions from cattle infected with bovine spongiform encephalopathy (variant CJD, sometimes referred to as the human equivalent to mad cow disease) or are secondary to receiving contaminated cadaver-derived growth hormone, dura mater graft, corneal transplants or through neurosurgical contamination. It is important to note that acquired prion diseases occur under very specific circumstances.

Prion disease is not transmissible from person to person through normal contact or through environmental contamination. It is not spread through airborne droplets, as is tuberculosis, nor by blood, as are hepatitis and human immunodeficiency virus.

The average worldwide occurrence of prion disease is roughly one to two cases per million people per year. This number is consistent globally among countries with surveillance centers. Despite being referred to as a "one in a million" disease (i.e., the incidence rate), the CDC estimates that approximately 1 out of every 6,000 deaths in the United States is related to a prion disease each year. This is due to the fact that age is a risk factor for developing the disease and of the variation of illness duration. Given this number, it is highly likely that a mortuary professional has served a family whose loved one died from a diagnosed or even undiagnosed prion disease. Within the scientific community, prion disease is known as the perfect "mimicker" of other well-known neurodegenerative diseases, including Alzheimer's, Parkinson's, Lewy Body Dementia, etc., due to the overlap of symptoms.

Prion disease is a rapidly progressive neurodegenerative disorder. There is currently no cure or treatment, and it is, unfortunately, always fatal. The majority of patients die within one year of disease onset, with a mean duration of four to six months. Treatment focuses on minimizing symptoms and maintaining quality of life for both patients and their family/caregivers.

Once a diagnosis of prion disease is made, it is recommended that end-of-life concerns be addressed quickly. Hospice care is usually brought in, and it is recommended that families make pre-arrangements with a funeral home, including whether the family wishes for a postmortem examination before cremation/embalming.

The National Prion Disease Pathology Surveillance Center (NPDPSC) was established in 1997 in the Department of Pathology at Case Western Reserve University. Its purpose is to acquire tissue samples and clinical information from as many cases of human prion disease occurring in the United States as possible in order to help monitor the possible occurrence of variant CJD and other emerging prion diseases in the country. It also assists clinicians in making a clinical diagnosis of prion disease by offering a variety of tests, including cerebrospinal fluid and genetic testing.

Although the United States has seen only a few cases of bovine spongiform encephalopathy in our cattle herds, chronic wasting disease (CWD), a prion disease of elk and deer in both free range and captive herds, has been spreading across North America over the past few decades. Since there are an estimated 22 million elk and deer in the United States and a large number of hunters who consume elk and deer meat, many humans could be potentially exposed to CWD. To date, there have been no reported cases of CWD infection in humans.

The NPDPSC coordinates with the Centers for Disease Control and Prevention, as well as with individual state health departments to monitor cases from CWD-endemic areas. Furthermore, it is doing experimental research on CWD transmissibility using animal models. Continued surveillance is necessary to investigate whether CWD or other novel prion diseases can be transmitted to people.

Currently, the only way to confirm a diagnosis of prion disease is through autopsy and the examination and characterization of formalin-fixed and frozen brain tissue. With funding from the CDC, NPDPSC offers a free-of-charge brain-only autopsy to families with loved ones suffering from suspected prion disease. At this time, the center is not aware of any state that requires an autopsy in cases of suspected prion disease/CJD. Enrollment in the program is voluntary and can be withdrawn at any time prior to autopsy. Families often choose to enroll their loved one in the program so they can confirm a diagnosis, determine the subtype and learn whether the disease is sporadic or genetic in etiology. The center covers all autopsy-related expenses, including but not limited to round-trip transportation between the storage and autopsy site (as needed), testing for prion disease and additional miscellaneous costs.

The center's network of public and private autopsy providers across the country works closely with mortuary professionals to make arrangements and provide education regarding the disease. It is the responsibility of the contracted mortuary professional to remove a decedent from their place of death. For patients enrolled into our autopsy program, the center requests that the body is refrigerated within four hours of death or as soon as possible. A body can be safely and discreetly removed using appropriate standard infection control measures, including personal protective equipment. No special precautions are required.

Patients enrolled in the center's autopsy program are cared for with dignity. Its staff and licensed mortician take painstaking care to ensure that the level of transmissibility of prion disease is at a minimum.

Misinformation circulated regarding prion disease can be a considerable barrier affecting the care and compassion that is extended to families. Education is difficult for numerous reasons. Given that prion disease is rare, it is not a disease with which many mortuary professionals are familiar. Misinformation about how to handle the decedent and the transmissibility of the disease has often resulted in families being denied services such as embalming. Per the CDC, embalming bodies of CJD patients who have been autopsied can be safely performed using standard infection control measures as outlined in the protocol "Infection Control Guidelines for Transmissible Spongiform Encephalopathies," distributed by the World Health Organization (WHO). An open-casket viewing should not be prohibited. However, if an autopsy has been performed, it is recommended that family members be advised to avoid superficial contact with the body (such as touching or kissing the decedent's face).

There are also no special burial or cremation requirements. Interment of bodies in closed caskets presents no significant environmental contamination risk, and cremated remains can be considered sterile and safe as the

LEARN MORE

To increase your knowledge about prion diseases, also check out these resources:

- cdc.gov/prions/cjd/funeral-directors.html
- cjdsurveillance.com
- cjdfoundation.org

infection agent does not survive cremation temperatures. According to the aforementioned WHO guidelines, any mortuary working surfaces that accidentally become contaminated with a potential CJD agent (e.g., spinal fluid, brain tissue, etc.) should be flooded with sodium hydroxide or bleach, left undisturbed for at least one hour, then (using gloves) mopped up with absorbent disposable rags and surfaces swabbed with sufficient water to remove any residual disinfectant solution. The NPDPSC serves as a resource for funeral homes and embalmers; its autopsy team is available to answer any questions and can be reached at 216-368-0587.

Family advocacy organizations, such as the Creutzfeldt-Jakob Disease Foundation (CJDF) in the United States, are a tremendous resource for families, mortuary professionals and the scientific community. Trained staff and volunteers are available seven days a week to answer calls from family members, physicians, hospice and healthcare providers, mortuary professionals and others. Over the span of several years, the center has partnered with CJDF to provide education and support to mortuary professionals regarding servicing families with suspected prion disease. Our autopsy team, along with our licensed funeral director and mortician, has participated in continuing education seminars for the Embalmers Association of Cleveland, the Alabama Funeral Directors Association, the Funeral Directors Association of the Falls Cities and the annual conference of the Oregon Funeral Directors Association.

It is our goal that through our various outreach efforts and continuing education seminars, we can assist and educate funeral and mortuary professionals in the appropriate handling of prion disease patients. We also aim to clear up any misconceptions or misinformation about the disease that may be prohibiting the use of a facility due to a prion disease diagnosis and the family's wishes of an autopsy to confirm such a diagnosis.

We hope the information presented has enriched your understanding of prion diseases and invite you to contact our autopsy coordinators at any time for additional information or guidance.

Meghan Lewis is autopsy specialist, Katie Glisic is center coordinator, Danielle Jordan is autopsy program manager and Dr. Brian Appleby is director of the National Prion Disease Pathology Surveillance Center.