Sporadic Creutzfeldt-Jakob Disease (sCJD)

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[Ted Pestorius] Welcome to this CDC podcast. I'm your host, Ted Pestorius. Today, I'm talking to Dr. Lynne Sehulster, a microbiologist with CDC's Division of Healthcare Quality Promotion. We're discussing an article on sporadic Creutzfeldt-Jakob disease published in the February 2009 issue of Emerging Infectious Diseases. Welcome to the show, Lynne.

[Lynne Sehulster] Well, thanks for having me.

[Ted Pestorius] Lynne, tell us a little about sporadic Creutzfeldt-Jakob disease.

[Lynne Sehulster] Creutzfeldt-Jakob disease, or CJD, is a rare neurodegenerative disease. This disease is caused by a pathological accumulation in the brain of an abnormal protein known as prions. The disease is characterized by dementia, which includes cognitive impairment, as well as profound motor skills impairment. CJD is uniformly fatal and is usually attributed to pneumonia associated with dementia. As the disease progresses over a course of 6 to 12 months, the patient loses neurologic function, which includes impaired cognitive ability, memory, and recognition. The symptoms of dementia from CJD may sometimes be similar to those that occur with Alzheimer's disease, but its gross neuropathology is different from that seen in an Alzheimer's patient. Also, the CJD patient's deterioration is more rapid. CJD affects the patient's motor skills, including loss in balance and jerky movements when the patient is surprised or startled. People who have CJD become increasingly nonresponsive to stimuli, and they may become mute shortly before death.

[Ted Pestorius] Lynne, what are the different types of CJD?

[Lynne Sehulster] There are three categories of CJD in the United States—familial, sporadic, and iatrogenic. Familial CJD is associated with certain inheritable genetic traits. Sporadic CJD appears to occur randomly with no apparent exposure event. Of all the CJD cases in the United States, the sporadic form is the most prevalent, accounting for about 90 percent of cases. Sporadic CJD is a disease that primarily affects elderly people in the United States and abroad. CJD patients typically range in age from their late 50s and older. The greatest number of cases occur in people in their 70s. Iatrogenic CJD cases have been associated with exposure to CJD-infected material during neurosurgical procedures, pituitary-derived hormone therapy, or neurologic tissue transplants. Fortunately, both the iatrogenic form and familial form of CJD are very rare in this country.

[Ted Pestorius] Tell us about the article.

[Lynne Sehulster] In this article, the authors were trying to determine trends of CJD infections in Japan by surveying past patient registries for any unidentified risk factors of patient risk for CJD. The authors didn't find any predictable patterns. They reviewed some infection control precautions doctors might take if a patient had potential exposure in a previous or upcoming

surgery. Their options may include using disposable instruments in these circumstances, which is one of the recommended strategies according to the World Health Organization's guidelines for preventing infection with prion diseases, such as CJD. In addition, doctors can look for any medical or diagnostic events or trends in a patient's history that could alert health care professionals to risk. Later, if that person should need surgery, particularly neurosurgery, then medical professionals would manage the patient and medical instruments accordingly. Health care professionals can take a proactive approach to medical instrument management.

[Ted Pestorius] Thanks, Lynne. Why are we hearing so much more about CJD now?

[Lynne Sehulster] Scientists are beginning to learn more about CJD through sophisticated molecular biology techniques. More research is being done to understand its natural history and to explore possible new treatments. We know that prions in neurologic tissue can lead to CJD, but we're now identifying prions in other tissues and organ systems. We've yet to determine what impact this discovery will have on CJD epidemiology. As we learn more about the biochemistry of CJD, we're trying to understand how prions are created. Prions that cause CJD are an abnormal form of normal cellular proteins, and scientists are trying to find out how the change in conformation occurs and what triggers this change. Learning more about the genetics of the disease is valuable research. The existence of familial CJD suggests there might be a genetic predisposition for CJD, whereas sporadic CJD might result from a randomly occurring mutation in the gene that produces the normal version of the prion protein.

[Ted Pestorius] What can people do to protect themselves and their families and communities from sporadic CJD here in the United States and abroad?

[Lynne Sehulster] Unfortunately, there's not much. There's no prevention, treatment, early diagnosis, or vaccine for sporadic CJD. Some possible prevention activities for iatrogenic CJD can include networking by neurologists, neurosurgeons, and nurses to see if they have a suspect case of CJD among their patients. Then, the infection control team can manage the medical instruments according to the recommendations of CDC and the World Health Organization. Being aware of signs and symptoms of early CJD is especially important.

[Ted Pestorius] What's the public health importance of this study?

[Lynne Sehulster] We're looking at ways to identify reliable predictors and risk factors to help us be more proactive in identifying sporadic CJD cases. As I mentioned earlier, it's possible that the sporadic cases might be caused by a random mutation of the normal prion gene; however, the result of this mutation may take a long time to manifest, and we don't have a feel for how long it takes to build up a critical level of abnormal prions in the brain. We do know that with iatrogenic CJD associated with neurosurgery, the disease can occur approximately 2 years after the exposure event. Although there's no proven intervention for CJD once a bona fide exposure has occurred, health care professionals can take steps to reduce the potential for transmission by being more alert to family history of dementia and neurological signs and symptoms suggestive of dementia and motor skill impairment. We encourage the health care community to become more aware of the disease.

[Ted Pestorius] This discussion with Dr. Lynne Sehulster was prompted by an article in the February 2009 issue of Emerging Infectious Diseases. These articles, and others on emerging bacterial and viral diseases, can be read online at www.cdc.gov/eid. You can submit your comments on this interview to eideditor@cdc.gov. For Emerging Infectious Diseases, I'm Ted Pestorius.

[Announcer] For the most accurate health information, visit www.cdc.gov or call 1-800-CDC-Info 24/7.