



CONGRESSIONAL REQUESTS

Creutzfeldt-Jakob Disease Foundation Advocacy Visits

FY2023

Overview

Creutzfeldt-Jakob Disease (CJD), is a rare, 100% fatal, degenerative brain disease that causes rapidly progressive dementia. CJD is transmissible and presently has no treatment or cure. Approximately 1 in 6,200 individuals will die from this disease in their lifetime; however, the unreported and undiagnosed number of cases remains unclear.

CJD surveillance receives modest support through the Centers for Disease Control and Prevention (CDC). We need your support to strengthen and continue the coordination of CJD and other prion disease surveillance activities and to assure the safety of the American public.

Variant CJD (vCJD), and Bovine Spongiform Encephalopathy (BSE)

One form of this disease in humans, variant CJD (vCJD), is known to be caused by ingesting beef contaminated with Bovine Spongiform Encephalopathy (BSE), commonly known as “mad cow” disease. The most recent U.S. case of variant CJD was announced in 2013 and confirmed by the National Prion Disease Pathology Surveillance Center (NPDPSC) in 2014. Limited BSE testing by the USDA adds another layer to the already deepening concerns regarding possible risks to humans. In recent years, the USDA has decreased random testing for BSE from 40,000 to 25,000 tests per year (12,719 tests in 6 months, or 1 test per 3,302 live cows). Hence, surveillance of BSE in this country is largely dependent on demonstrating the lack of transmission to humans through human disease surveillance. The vCJD case identified by NPDPSC in 2014 exemplifies the persistent risk for vCJD acquired in unsuspected geographic locations and highlights the need for continuing prion disease surveillance and awareness to prevent further dissemination of vCJD. The two most recent cases of vCJD in Europe are believed to be due to occupational exposure and several cases of vCJD have been transmitted between individuals via blood transfusions. Hence, vCJD risk is not confined to eating contaminated food.

Chronic Wasting Disease (CWD)

Emerging laboratory data show that Chronic Wasting Disease (CWD), a naturally occurring prion disease of deer and elk, could potentially transmit to humans and other mammals, posing a new threat to public health. Human surveillance through brain tissue examination is the only way to definitively diagnose human prion diseases, determine their origin, and determine whether the spread of CWD found in elk and deer in 29 states in the U.S. and in 4 Canadian provinces has become a human risk. A study in progress has reported that CWD was transmitted to macaques (primates that are genetically similar to humans) by feeding them contaminated deer meat. Unlike the BSE outbreak in cattle, CWD prions are highly infectious among its own species and the disease transmits by contact and through contaminated environment, including soil and plants, in free ranging and farmed animals. Additionally, multiple lines of experimental evidence indicate that sheep and cows are susceptible to CWD. Since CWD has been proven to cross several species barriers, this opens up the possibility of

transmission to humans as well, either directly by eating contaminated venison or indirectly through infected domestic animals. Continued prion disease surveillance, particularly through examination of human brain tissue, is imperative to evaluate whether CWD has or can spread to humans.

The NPDPS, funded by the CDC and located at Case Western Reserve University in Cleveland, Ohio, is our line of defense against the possibility of an undetected U.S. human prion disease epidemic as experienced in the United Kingdom.

Prion disease surveillance is funded at \$6.5 million/year. That figure has increased by just \$500,000 over the past seven years, despite increasing costs of surveillance. Expenses have since risen for the resources required to perform adequate surveillance such as increasing number of cases as expected by the aging American population, increasing autopsy costs over time, screening for COVID19, and taking extra precautions necessary for COVID19. Without an increase in funding commensurate with these increased expenses, surveillance will be compromised.

Key Request 1: We ask for Congressional support in increasing prion disease surveillance's appropriation by \$1 million, for a total of \$7.5 million. This would allow the NPDPS to meet increasing autopsy costs and continue to develop more efficient detection methods while providing an acceptable level of prion surveillance. Reduction of funding or maintaining static funding to the NPDPS would eliminate an important safety net to U.S. public health, making the U.S. the only industrialized country lacking prion surveillance, which in turn would jeopardize the export of U.S. beef. The increase in funding would allow the NPDPS to expand its scope to address the growth in CWD among deer and elk, and explore whether CWD could spread to humans. Additionally, increasing prion disease surveillance in the U.S. increases surveillance at the national (CDC) and state (state public health departments) levels, which has been severely affected by competing concerns within the CDC division (e.g., COVID19).

Background:

The NPDPS is funded entirely by the CDC from funds allocated by Congress. The CDC traditionally keeps approximately half of the appropriation for epidemiologic surveillance projects and funding prion disease surveillance at the state level.

Increasing the appropriation from \$6.5M to \$7.5M will allow the NPDPS to persist and continue to develop more efficient detection methods while providing an acceptable level of prion disease surveillance. Acceptable national surveillance is not possible at a lower level of funding. The requested \$1M addition to the appropriation (total of \$7.5M) would enable the NPDPS to maintain appropriate surveillance, tissue collection, diagnostics and diagnostic test development of prion disease cases from CWD endemic states to determine whether CWD is transmissible to humans and if so, to what extent this poses to public health (e.g., transmission risks from human to human).

The National Prion Disease Pathology Surveillance Center is the only laboratory-based organization in the U.S. that monitors human prion diseases and is able to determine whether a patient acquired the disease through the consumption of prion contaminated beef ("mad cow" disease) or meat from elk and deer affected by chronic wasting disease (CWD).

The NPDPS also monitors all cases in which a prion disease might have been acquired by infected blood transfusion, from the use of contaminated surgical instruments, or from contaminated human growth hormone. Because standard hospital sterilization procedures do not completely inactivate prions that transmit the disease, these incidents put a number of patients under unnecessary risk and require costly replacement of contaminated surgical equipment.

The NPDPSC also plays a decisive role in resolving suspected cases or clusters of cases of food-acquired and medically transmitted prion disease that are often magnified by the media, stirring intense public alarm. To date, the NPDPSC has examined approximately 8,000 suspected cases of suspected prion disease and has definitely confirmed presence and type of prion disease in more than 4,800 cases.

The NPDPSC is the primary line of defense in safeguarding U.S. public health against prion diseases because the U.S., unlike other BSE affected countries such as the UK, the European Union, and Japan, does not have a sufficiently robust animal prion disease surveillance system.

The NPDPSC offers assurances to countries that import (or are considering importing) meat from the United States, that the U.S. is free of indigenous human cases of “mad cow” disease. In the past, South Korean and Chinese health officials resumed importation of U.S. beef to their country after a visit to the NPDPSC provided assurances regarding rigorous human prion surveillance.

Since its inception in 1997, the NPDPSC has collected and stored approximately 8,000 brains and many more samples of cerebrospinal fluid from cases of suspected prion disease, making it the largest prion disease biobank in the world. Increased funding is required to continue to preserve these precious specimens for future international research efforts as well as to serve as reference materials to evaluate potential emerging prion diseases (e.g., chronic wasting disease).

Key Request 2: The CJD Foundation and families affected by CJD request that the NIH consider Prion Disease as an Alzheimer’s Disease Related Dementia (ADRD), which would allow NIH funds allocated to these groups of diseases to also be applied to prion diseases.

Background

Some funding for prion disease research comes from the NIH, primarily the National Institute of Neurological Disorders and Stroke (NINDS). However, the CJD Foundation, prion disease and other ADRD researchers, and family members recognize the need to devote more funds to exploring prion disease, and especially to integrating the scientific discoveries of prion disease into other ADRDs in order to improve diagnosis, biomarkers, and develop treatments.

There are many similarities between ADRD and prion disease. Primarily, all ADRD are due to an abnormal form of normal cellular protein that causes disease (e.g., a-beta and tau in Alzheimer’s disease, alpha-synuclein in Lewy body dementia). These abnormal proteins affect certain areas of the brain causing neurodegeneration and neurocognitive decline. In the same way, prion disease is due to an abnormal form of the normal prion protein. For several years, ADRD researchers have exploited initial discoveries in prion disease in order to advance the ADRD field. As with prion disease, proteins implicated in ADRD can be transmitted between cells. These abnormal proteins can also be amplified using protein amplification techniques initially created to study prion disease. Hence, ADRDs have already benefited from prion disease research and will undoubtedly continue to do so. Inclusion of prion disease as an ADRD:

1. Allows ADRD researchers to study the prion paradigm in other related conditions which in turn enables the use of more valid animal models as animals naturally develop prion disease but not other neurodegenerative diseases.
2. Allows technologies originally developed for prion disease to be used in other ADRDs without the need for separate funding.
3. Allows potential treatment targets to be expanded across ADRD and prion disease.
4. Allows caregiver and health services research of ADRD to be applied to prion disease, which shares many of the same challenges.