Creutzfeldt-Jakob Disease: An Infectious Protein

DPH Recognizes November 12th as International Creutzfeldt-Jakob Disease Awareness Day with information about this disease

Creutzfeldt-Jakob Disease (CJD) is a rare, but fatal, neurodegenerative brain disorder within a group of illnesses called prion diseases. A prion is an abnormally shaped form of a normally harmless protein found in human brains. Prions can infect normal proteins causing them to misfold and form into prions. As prions replicate and infect new proteins, they grow within nerve cells, causing brain tissue to die. CJD was first recognized in the early 1920s by German neurologists Hans Gerhard Creutzfeldt and Alfons Maria Jakob.

There are different types of prion disease:

- **Sporadic CJD**
  - ~85% of all CJD cases are sporadic. This is the most prevalent form of CJD with 1 to 2 cases per 1 million population per year in the United States (actual numbers may be higher due to underreporting).
  - Cause is unknown.
- **Genetic CJD**
  - ~5-15% of all CJD are genetic.
  - Caused by a genetic mutation of the prion protein gene.
- **Variant CJD**
  - ~Less than 1% of all CJD cases. Four cases of, Variant Creutzfeldt–Jakob disease (vCJD), have been reported from the United States (actual numbers may be higher due to underreporting).
  - Caused by eating beef contaminated with prion or by receiving blood transfusions from persons with variant CJD. Note: variant CJD is not the same as mad cow disease.
- **Iatrogenic CJD**
  - ~Less than 1% of all CJD cases.
  - Caused by medical equipment or transplant organs contaminated by prions.

Both men and women are equally affected by CJD, and this disease commonly occurs in people between the ages 40 and 70. The onset of CJD is usually characterized by failing memory, behavior changes, impaired coordination, and visual problems. The illness progresses as dementia, blindness, weakness of extremities, and involuntary movements. Sadly, there is no known cure for CJD, and the disease is usually fatal within a year of symptom onset.

On International Creutzfeldt-Jakob Disease Awareness Day, DPH encourages residents to learn more about CJD and to educate others on this illness. Additionally, you can wear purple to honor loved ones affected by CJD and to increase awareness!

For more information about prion diseases and CJD, visit: https://dhss.delaware.gov/dhss/dph/epi/cjd.html, www.cjdfoundation.org, or https://www.cdc.gov/prions/index.html