Creutzfeldt-Jakob’s Disease, classic (sporadic)

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Etiological Agent:
Misshapen prion protein - PRP\textsuperscript{sc} (7).

Transmission:
Transmission primarily occurs three different ways:
1. Sporadic – Randomly without explanation (3,4)
2. Inherited – Genetic mutation passed through families (3,4)
3. Contamination – Exposure to infected human tissue during medical procedures (iatrogenic). Exposure from eating infected beef (variant) (3,4)

Reservoirs:
Elderly Humans (median age 68) or ingestion of infected beef (median age 28) (1,6).

General Characteristics of MO:
Self-replicating protein called a prion. The gene that expresses the prion, in humans, can be found on the 20\textsuperscript{th} chromosome. Expression of the prion occurs naturally in non-infected humans. The non-pathogenic prion, however, can become rearranged creating a chain reaction, leading to infection (7).

Key Tests for identification:
Identification of an individual infected with Creutzfeldt-Jakob’s Disease can only be confirmed by an autopsy of the individual’s brain, post mortem. Neurological tests such as an MRI, electroencephalograms, and spinal taps are often used to rule out other neurological disorders (2,5).

Signs and Symptoms:
The prion that causes Creutzfeldt-Jakob’s Disease may incubate within an individual for as long as 50 years. Within the first couple of months of showing signs, individuals with Creutzfeldt-Jakob’s Disease experience memory loss and changes in personality. (2) Within the oncoming months after initial symptoms, the individual will rapidly lose mental capacities and become comatose. (3) Death usually occurs, within a year of onset of symptoms, due to respiratory or cardiac failure (3).

Historical Factors:
Creutzfeldt-Jakob’s Disease was first identified in the 1920’s by Hans Creutzfeldt and later by Alfons Jakob. (1) Prions were proposed as the primary etiological agent for Creutzfeldt-Jakob’s Disease in 1982 by researcher Stanley Prusiner. (12) In 1996 researchers at the *Eunice Kennedy Shriver* National Institute of Child Health and Human Development (NICHD) found that the misshapen prions, in individuals infected with Creutzfeldt-Jakob’s Disease, bonded to Mahogunin in the individual’s brain, leading to brain degeneration. (8)

**Virulence Mechanisms:**
Normal prion (PRP<sup>c</sup>) becomes misshapen and turns into prion (PRP<sup>sc</sup>). The PRP<sup>sc</sup> prion becomes a template to misshapen other PRP<sup>c</sup> prions. PRP<sup>sc</sup> goes on to accumulate in the brain of the infected host, causing degeneration of brain tissue. PRP<sup>sc</sup> binds with a protein found in brain cells called Mahogunin. This binding process denies brain cells access to Mahogunin, causing them to die. This ultimately, on a larger scale, causes abscesses in the brain (8).

**Control/Treatment:**
Classic or sporadic Creutzfeldt-Jakob Disease occurs randomly and infects the individuals nervous tissue, making it usually not contagious (4,6). There is no effective treatment for Creutzfeldt-Jakob’s Disease. Treatment of pain is usually recommended (4,5). Medical instruments that come in contact with suspected infected tissue should be considered contaminated and incinerated (7).

**Prevention/Vaccine info, new trials?**
Organ donation and donating of blood of an individual infected with Creutzfeldt-Jakob’s Disease should be prevented (6). There are no known vaccines for Creutzfeldt-Jakob’s Disease. Research is currently being conducted by the Eunice Kennedy Shriver National Institute of Child Health and Human Development (NICHD) to contain misshapen prions in infected individuals and possible ways to restore Mahogunin, when it is depleted in an infected individual (8).

**Local cases or outbreaks:**
There have been no reported cases of Creutzfeldt-Jakob’s disease in Travis County from 2013 up until now. (11)
In 2012, there were 9 confirmed cases and 9 probable cases of sporadic Creutzfeldt-Jakob’s Disease reported in Texas. (9) The CDC has tracked reported cases of classic Creutzfeldt-Jakob’s Disease to be approximately 1-
2 cases per million a year, with the majority of the reported individuals being 60 years of age or older. (1)

**Global cases or outbreaks**
Incidence of Creutzfeldt-Jakob’s disease on a global scale remains rare. From 1996 to 2012, there was approximately 1 case per million reported. (10)

**References:**


