

Protein to Disease

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Mad Cow Disease / Bovine Spongiform Encephalopathy (BSE) and Creutzfeldt-Jakob Disease (CJD)

List of tasks:

- 1) Research points in need to know
- 2) Organize research into a storyboard
- 3) Make a video (whiteboard / vocal explanation)
- 4) Edit the video

Research:

Symptoms/Effects:

CJD: Problems with muscle coordination, impaired memory, judgement, and thinking, and impaired vision. Can cause seizures and spasms. Dementia develops in later stages of CJD. Fatal within 13 months of the onset of symptoms.

Treatments/Barriers to curing:

There is no current cure to CJD. No treatment can completely control the disease, but some opiate drugs are available to alleviate pain. Clonazepam (sedatives) and sodium valproate (anticonvulsants) are also used to relieve myoclonus (seizures).

Researchers at New York University School of Medicine recently took a crucial step in developing an effective treatment. They found 4 compounds that significantly delayed disease onset in mice. Because prion diseases are slow to develop, any treatment that delays their initial symptoms long enough could potentially be life saving.

Who it affects:

Onset of symptoms typically starts around age 60. Ninety percent of individuals die within a year. CJD is very rare, only occurring in about one in a million people.

How it is spreads:

This disease is spread to humans by eating diseased meat from a cow with Mad Cow disease. Anyone can get it. It can also be hereditary if you have a family history of the disease.

What is mad cow disease?

- Creutzfeldt-Jakob Disease (CJD), is a transmissible, slowly progressive, degenerative, and fatal disease affecting the central nervous system.
(<https://www.webmd.com/brain/mad-cow-disease-basics#1>)

- It is infectious and can be transmitted between people through injections or by consuming infected brain or nervous tissue. It can also be hereditary if you have a family history of CJD.

What are treatments, cure?

- There is no cure
- Treatments only to ease pain
- Researchers at NYU School of Medicine recently took a crucial step in developing an effective treatment. They found four compounds that significantly delayed disease onset in mice. Because prion diseases are extremely slow to develop, any treatment that delays their initial symptoms long enough could potentially be life saving. And two of these anti-prion compounds are drugs already used to treat other conditions in humans, meaning that clinical trials of them may soon to be constructed.

What protein is affected by mad cow disease?

- The infectious agent that causes CJD is an abnormal version of a protein normally found on cell surfaces, called a prion. For reasons still unknown, this protein becomes altered and destroys nervous system tissue, the brain and spinal cord.

Works Cited:

<https://www.ninds.nih.gov/Disorders/Patient-Caregiver-Education/Fact-Sheets/Creutzfeldt-Jakob-Disease-Fact-Sheet> - treatments, symptoms

<https://www.nhs.uk/conditions/creutzfeldt-jakob-disease-cjd/causes/> - proteins

<https://www.webmd.com/brain/mad-cow-disease-basics#1> - what it is

https://www.medicinenet.com/creutzfeldt-jakob_disease/article.htm - who

<https://en.m.wikipedia.org/wiki/Prion> - prion protein

What is a prion

Prions are misfolded proteins which characterize several fatal neurodegenerative diseases in animals and humans. It is not known what causes the normal prion protein to misfold; the abnormal 3-D structure is suspected to confer infectious properties. The word prion derives from "proteinaceous infectious particle" ([https://\(en.m.wikipedia.org/wiki/Prion\)](https://en.m.wikipedia.org/wiki/Prion))