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Alzheimer's on a Fast Track? A Comprehensive Look Into the Fatal World of CJD, Creutzfeldt Jakob Disease

Creutzfeldt Jakob disease, also known as CJD, is a rare neurodegenerative disorder of the brain that can lead to the development of dementia. CJD can commonly be classified as a prion disorder due to its abnormally assembled abundance of distorted prion glycoprotein in the brain (Kerr et al., 2018). Symptoms of the disease include drastic changes to an individual's personality and thinking, in addition to a deficiency in memory, vision, vocalizations, or coordination (Mayo Clinic Staff, 2023). Difficulty in food ingestion may also be a concern coupled with an accumulation of insomnia and acute muscle gestures. It has been determined that symptoms of the disease are prone to worsen within the following weeks and months ahead of diagnosis. The prognosis for the disease is extremely poor with an expected maximum one-year life expectancy following diagnosis. Once the disease has progressed enough, individuals have a high risk of falling into a coma until they reach death.

In 1920, Alfons Maria Jakob, a well-known German neuropathologist, had suspected a non-traditional neurodegenerative form of dementia in six individuals he had studied. Following this in 1921, Hans Gerhardt Cruetzfelt, who had been another German neuropathologist submitted similar case findings to that of Jakob. Thus the discovery of such a disease was credited to both scientists and in 1922 began a term called the "Creutzfeldt Jakob disease" (Henry & Murphy, 2017).

Diagnosis of the disease commenced in all parts of the world rarely, but cases were primarily seen in the United Kingdom in the twentieth century. While the prevalence of the disease remains primarily in middle-aged adults, the risk of concurring with the disease poses a larger risk with aging. The risk factor for an average adult for accumulating CJD is one in every one million adults, while between the years of 2016 and 2020 in the United States, case prevalence has risen to a risk factor of five in every one million adults over the age of 55. The latest death toll results globally from the disease have resulted in over 500 individual deaths (Kerr et al., 2018).

Confirmation of diagnosis for CJD includes a brain biopsy, symptoms of the presence of progressive neurodegeneration, MRI, or an EEG. These demonstrated tests include the detection of the 14-3-3 protein as well as an abnormal prion protein collection (Zerr, 2022). Qualitative assay techniques are used through EEG and MRI exams for optimal accuracy and sensitivity to tissue. This RT-QuIC assay technique was developed by the National Prion Disease Pathology Surveillance Center in 2015 with high rates of efficiency.

There are several distinctive types of CJD. The first and most common accounting for 85% of documented cases has been proven to be sporadic, entailing that the disease arose spontaneously. This is frequent in individuals between the ages of 55 to 75 and typically has a survival rate of between 4 to 8 months post-diagnosis. The second type of CJD is genetic and or familial which accounts for 15% of cases. This may be detected through the presence of a mutation PRNP gene within the prion protein. The third type of CJD accounting for less than 1% of cases is acquired through transmission from other humans or animals. This occurs through some sort of exposure to the infected nervous and or brain tissue that can be detected in an MRI scan, and is prevalent in primarily young adults. Lastly, there is the variant type of CJD, acquired through ingestion of infected beef which is more prevalent in the United Kingdom and or France (Iwasaki, 2016).

Recent studies have yet to determine a definite cure for the disease and with this, the primary focus remains on the treatment of symptoms for the infected patients. Antidepressant medications have been frequently used for these reasons to minimize pain and anxiety for the suffering individual (NHS, 2021). Preventative measures for accumulating CJD remain as the prevention for individuals with first-degree family members from participating in blood donations in addition to recommendations for individuals who consume deer or elk meat to subject it to genetic testing before consumption (Iwasaki, 2016)

A new biomedical invention of a monoclonal antibody targeted at the brain labeled as PRN100 began experimental testing on six patients in the years 2018 and 2019. Although all six patients eventually died as a result of CJD, the treatment was demonstrated to stabilize symptoms in three of these patients. Researchers are expecting further studies in the near future with larger patient sample sizes as well as improvements to the treatment in order to give conclusive final data on the efficiency of the treatment (NIHR, 2022)

It is imperative for future studies to concern the Creutzfeldt Jakob disease in their research to find an efficient and or conclusive treatment. Awareness of the disease should be prevalent throughout communities as well as education systems in order to instill a drive to create fundraisers and other sorts of events to aid in research progression.

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