Journal of
Neuroinfectious Diseases
ISSN: 2314-7326

Zhao, J Neuroinfect Dis 2025, 16:2

Mini Review Open Access

Creutzfeldt-Jakob Disease (CJD): A Rare but Fatal Brain Disorder

Zhao Ming*

Department of Neurology, Huazhong University of Science and Technology, China

Introduction

Creutzfeldt-Jakob Disease (CJD) is a rare, degenerative, and fatal brain disorder affecting approximately one in a million people worldwide each year. It belongs to a group of conditions known as prion diseases, which are caused by abnormal proteins called prions that damage brain tissue. CJD progresses rapidly, leading to severe neurological decline and ultimately death, usually within a year of symptom onset. Though rare, CJD has drawn significant attention due to its devastating effects and its association with variant forms linked to contaminated meat. This article explores the causes, symptoms, types, diagnosis, treatment, and ongoing research into CJD. Creutzfeldt-Jakob Disease (CJD) is a rare but fatal neurodegenerative disorder that affects the brain, leading to rapid cognitive decline and severe neurological impairment. It is classified as a prion disease, caused by the accumulation of abnormally folded proteins called prions. These infectious proteins trigger a chain reaction, converting normal proteins into the misfolded form, leading to progressive brain damage. Unlike bacterial or viral infections, prion diseases do not trigger an immune response, making them difficult to detect and treat. CJD has an incidence rate of approximately one in a million people per year, though certain genetic and environmental factors can increase the risk. The disease primarily affects older adults, typically appearing in individuals between the ages of 55 and 75. However, variant forms of CJD, linked to consuming contaminated beef from cattle with bovine spongiform encephalopathy (BSE or "mad cow disease"), have been reported in younger individuals [1,2]. The symptoms of CJD develop rapidly, starting with memory loss, confusion, and personality changes before progressing to severe dementia, involuntary muscle movements, vision problems, and loss of coordination. Due to its aggressive nature, most patients succumb to the disease within a year of symptom onset. Currently, there is no cure for CJD, and treatment is limited to palliative care aimed at alleviating symptoms. Research is ongoing to develop early diagnostic methods and potential therapeutic strategies to slow disease progression [3].

Discussion

Creutzfeldt-Jakob Disease (CJD) is a devastating neurodegenerative disorder that progresses rapidly, leading to severe cognitive and motor impairment. As a prion disease, it differs from other brain disorders because it is caused by misfolded proteins, rather than bacteria or viruses. These abnormal prions induce a cascade of protein misfolding, leading to brain tissue deterioration, memory loss, and ultimately, death. Unlike many neurological diseases, CJD progresses aggressively, with most patients surviving less than a year after symptom onset. One of the major challenges with CJD is its diagnosis. Since symptoms overlap with other neurodegenerative diseases like Alzheimer's or Parkinson's, misdiagnosis is common [4]. Currently, diagnostic methods such as MRI scans, cerebrospinal fluid tests, and electroencephalograms (EEGs) can indicate CJD, but a definitive diagnosis is only possible post-mortem through brain tissue analysis. This highlights the urgent need for more advanced biomarkers and early detection techniques. Another significant concern is the lack of treatment options. There are no known therapies that can halt or reverse the progression of CJD. Management is purely supportive, focusing on pain relief, symptom management, and providing comfort care. Research into potential treatments, such as prion-targeting drugs and immunotherapy, is ongoing, but no breakthrough has been achieved yet. Prevention strategies mainly focus on reducing transmission risks for variant and iatrogenic CJD. Strict regulations in the meat industry, improved sterilization of medical instruments, and screening measures for blood donations have been implemented to minimize exposure to infectious prions. Overall, while CJD remains incurable, continued research into early detection, prion biology, and potential treatments offers hope for better management in the future. Raising awareness and supporting affected individuals and their families is crucial in dealing with this fatal condition [5,6].

Causes and Mechanism

CJD is caused by prions—misfolded proteins that trigger normal proteins in the brain to misfold and accumulate, leading to brain tissue damage and deterioration. Unlike bacteria or viruses, prions do not contain genetic material and are resistant to conventional sterilization methods, making them particularly challenging to control. The abnormal prions cause spongiform changes in the brain, creating holes in the tissue and leading to rapid neurological decline [7].

Types of Creutzfeldt-Jakob Diseas

Familial CJD (fCJD)

Inherited form caused by mutations in the PRNP gene, which encodes for the prion protein.

Accounts for 10-15% of all cases.

Often follows an autosomal dominant inheritance pattern, meaning a child of an affected parent has a 50% chance of inheriting the mutated gene.

Iatrogenic CJD (iCJD)

Transmitted through contaminated medical procedures, such as improperly sterilized surgical instruments or corneal transplants [8].

***Corresponding author:** Zhao Ming, Department of Neurology, Huazhong University of Science and Technology, China, E-mail: zhao@ming.cn

Received: 03-Mar-2025, Manuscript No: JNID-25-162519, **Editor Assigned:** 07-Mar-2025, Pre QC No: JNID-25-162519 (PQ), **Reviewed:** 18-Mar-2025, QC No: JNID-25-162519, **Revised:** 22-Mar-2025, Manuscript No: JNID-25-162519 (R), **Published:** 29-Mar-22025, DOI: 10.4172/2314-7326.1000554

Citation: Zhao M (2025) Creutzfeldt-Jakob Disease (CJD): A Rare but Fatal Brain Disorder. J Neuroinfect Dis 16: 554.

Rare but has occurred in cases involving human growth hormone treatments derived from infected cadavers.

Diagnosis

There is no single test to definitively diagnose CJD in a living person. Instead, doctors rely on a combination of clinical assessments, imaging tests, and cerebrospinal fluid analysis [9].

Electroencephalogram (EEG): Identifies characteristic brain wave patterns associated with CJD.

Magnetic Resonance Imaging (MRI): Detects specific brain abnormalities linked to prion disease.

Cerebrospinal Fluid (CSF) Tests: Looks for biomarkers such as 14-3-3 protein, which may indicate CJD.

Genetic Testing: Identifies mutations in the PRNP gene in suspected familial cases [10].

Brain Biopsy or Autopsy: The only definitive way to confirm CJD, but rarely performed before death due to risks.

Research and Future Directions

Scientists are actively researching potential treatments and early detection methods for CJD. Some promising areas of study include:

Antiprion Therapies: Drugs that prevent prion proteins from misfolding and accumulating.

Immunotherapy: Investigating whether the immune system can target and neutralize prions.

Gene Therapy: Exploring ways to modify or suppress the PRNP gene to prevent prion diseases.

Early Biomarkers: Developing better diagnostic tests for detecting CJD before symptoms appear.

Conclusion

Creutzfeldt-Jakob Disease is a devastating and incurable neurodegenerative disorder caused by misfolded prion proteins. Though rare, its rapid progression and fatal outcome make it a significant concern in neurology and public health. While no cure currently exists, ongoing research aims to improve early detection and develop potential therapies. Preventive measures, particularly in controlling variant and iatrogenic cases, remain crucial in reducing the incidence of CJD. Raising awareness and supporting affected individuals and families is essential as science continues to search for answers to this mysterious and deadly disease.

References

1. Parkinson J (2002) An essay on the shaking palsy. J Neuropsychiatry Clin Neurosci 14: 223-236.
2. Morris AD, Rose FC, Birkhauser, Boston, MA (1989) James Parkinson His Life and Times, Deaths associated with neurological conditions in England.
3. GBD 2015 Neurological Disorders Collaborator Group (2017) Global, regional, and national burden of neurological disorders during 1990-2015: a systematic analysis for the Global Burden of Disease Study 2015. Lancet Neurol 16: 877-897.
4. Savica R, Grossardt BR, Bower JH, Ahlskog J, Rocca WA (2016) Time trends in the incidence of Parkinson disease. Neurol 73: 981-989.
5. Darweesh SKL, Koudstaal PJ, Stricker BH, Hofman A, Ikram MA (2016) Trends in the Incidence of Parkinson Disease in the General Population: The Rotterdam Study. Am J Epidemiol 183:1018-1026.
6. Akushevich I, Kravchenko J, Ukraintseva S, Arbeev K, Yashin AI (2013) Time trends of incidence of age-associated diseases in the US elderly population: Medicare-based analysis. Age Ageing 42: 494-500.
7. Isotalo J, Vahlberg T, Kaasinen V (2017) Unchanged long term rural-to-urban incidence ratio of Parkinson's disease. Mov Disord 32: 474-475.
8. GBD 2016 Parkinson's disease Collaborators (2018) Global, regional, and national burden of Parkinson's disease in 1990-2016: a systematic analysis for the Global Burden of Disease Study 2016. Lancet Neurol 17: 939-953.
9. Savica R, Grossardt BR, Bower JH, Ahlskog JE, Boeve B, et al. (2017) Survival and causes of death among people with clinically diagnosed synucleinopathies with Parkinsonism: a population-based study. JAMA Neurol 74: 839-846.
10. Rossi A, Berger K, Chen H, Leslie D, Mailman RB, et al. (2018) Projection of the prevalence of Parkinson's disease in the coming decades: Revisited. Mov Disord 33: 156-159.