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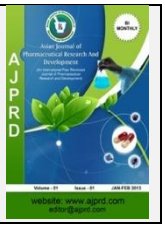

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Review Article

Creutzfeldt-Jakob Disease (CJD): A Deadly Neurodegenerative Condition; Its Epidemiology, Monitoring, Current Status of Prevention and Circumvention

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A B S T R A C T

Creutzfeldt-Jakob Disease CJDis a rare brain ailment that causes dementia. It is often referred to as Creutzfeldt-Jakob. It is a member of a class of illnesses called prion disorders that affect both humans and animals. Alzheimer's disease and Creutzfeldt-Jakob disease symptoms can be comparable.A rare yet deadly neurodegenerative condition is Creutzfeldt-Jakob disease (CJD). According to reports, it affects one in one to one and a half million people worldwide each year, with less than 1,000 cases occurring in the US.A deadly neurodegenerative condition, Creutzfeldt-Jakob disease (CJD) is also referred to as subacute spongiform encephalopathy or neurocognitive dysfunction caused by prion disease. Memory issues, behavioural changes, poor coordination, and visual impairments are some of the early indications. Dementia, uncontrollable movements, blindness, paralysis, and coma are later signs. Within a year, almost 70% of people pass away.In 1922, Walther Spielmeyer coined the term "Creutzfeldt–Jakob disease" in honour of the German neurologists Hans Gerhard Creutzfeldt and Alfons Maria Jakob. A protein called a prion folds abnormally to cause CJD. Misfolded proteins known as infectious prions can cause other misfolded proteins to do the same.People with CJD experience a variety of symptoms, such as uncontrollable muscle spasms or mobility issues, memory loss, and cognitive challenges. Because CJD causes so much damage, it is ultimately fatal.The diseases CJD and variant CJD (vCJD) are not the same, despite their very similar names. They're both prion illnesses. But eating meat from cows with bovine spongiform encephalopathy, also known as Mad Cow Disease, is linked to variant CJD. CJD mostly happens seldom and is sometimes referred to as "classic CJD" to prevent confusion.Although there isn't a known cure for Creutzfeldt-Jakob disease (CJD), the National Prion Clinic is doing clinical research to look at potential therapies. Currently, the goal of treatment is to keep the patient as comfortable as possible while using medications to lessen symptoms.The present state of Creutzfeldt-Jakob disease (CJD), its underlying causes, and possible treatments are assessed in this article.

Keywords: Creutzfeldt-Jakob disease (CJD), Epidemiology, Etiology, Diagnosis, Management

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INTRODUCTION

Prion proteins cause Creutzfeldt-Jakob disease (CJD), a rare, rapidly progressing, transmissible, and always fatal neurodegenerative illness. There is a lengthy incubation time for the disease. Hans Creutzfeldt initially described CJD in 1920, and Alfons Jakob followed in 1921 and 1923. Because the abbreviation "CJD" was more similar to his initials, Clearance J. Gibbs later began using the term "Creutzfeldt-Jacob disease."A rare and quickly progressing brain ailment, Creutzfeldt-Jakob disease (CJD) affects memory, reasoning, motor coordination, and brain tissue uniquely.An uncommon and deadly degenerative brain disease called Creutzfeldt-Jakob disease (CJD) is brought on by prion proteins. With an incidence of one case per million annually, this illness is part of a class of transmissible spongiform encephalopathies that impact people all over the world. In the United States, about 350 cases are diagnosed each year. Almost 70% of patients pass away within a year. Early and precise diagnosis of CJD affects overall results and therapeutic tactics.CJD usually manifests later in life, usually around age 60, and worsens rapidly. Approximately 70% of CJD patients pass away within a year. CJD cannot be contracted by touching an object, such as a doorknob, or being near someone with it. Nonetheless, physicians advise against donating blood, organs, or tissue if a patient has a

family history of CJD or has suspected or proven CJD. Prions are aberrant protein forms that cause CJD. Long sequences of amino acids make up proteins, which fold into specific shapes or conformations to support specific cellular functions. The neurological system contains the bulk of the normal, innocuous prion protein, however it is present throughout the body. Its overall function is not entirely clear. Normal prion proteins in the surrounding nerve cells are "transmitted" with this aberrant folding or conformation, which makes them infectious and transforms them into the form that causes the disease. The loss of nerve cells and other brain damage associated with CJD may result from aberrant prion proteins clumping together, or aggregating, once they have formed. CJD belongs to a family of human and animal diseases known as transmissible spongiform encephalopathies (TSEs) or prion diseases. The infective prion causes CJD and related disorders in people and TSEs in animals. Spongiform refers to the characteristic appearance of infected brains, which become filled with holes until they resemble sponges when examined under a microscope(1–4).The central nervous system (CNS) is the primary organ affected by CJD. The neuron, a special kind of cell with the ability to receive, store, and transfer information, is the primary functional unit of the central nervous system. Although certain parts of the brain can repair to a limited degree because of the presence of stem cells, central nervous system neurons cannot regenerate. The following characteristics set the central nervous system apart from other organ systems: blood flow autoregulation in the brain. Being protected by the cranium. Specific needs for metabolic substrates. Lack of a functional lymphatic system. Circulation of cerebrospinal fluid (CSF). Little immunologic monitoring. Different routes for tissue healing and damage response. The brain's neurons are arranged topographically, with functional domains found in areas that are physically defined. For instance, the hypothalamus is crucial for autonomic reactions, whereas the cerebral cortex governs voluntary movements. The somatotopic architecture of brain cells facilitates sensory and motor input from the central nervous system (CNS) to different parts of the body, which is useful for localizing neurologic abnormalities. Neurons vary in size and shape. Axons and dendrites, which vary widely in number from cell to cell, form synapses. The creation of proteins and neurotransmitters in nerve cells depends on Nissl bodies, which also vary in number. Neurofilaments contribute to nerve conduction and preserve the cytoskeleton.The cells that support neurons are called glia. Among them are the following: Star-shaped glial cells called astrocytes provide nourishment to neurons, serve as electrical insulators and nerve detoxifiers, shield the central nervous system from dangerous macromolecules, and aid in scar formation and CNS healing. The CNS axons are myelinated by oligodendrocytes. The ependymal cells that line the ventricles regulate the flow and generation of CSF. Macrophages in the central nervous system are made up of microglia. Brain neurons are damaged by CJD neuronal inclusions, which cause neurologic abnormalities, including myoclonus, in the latter stages of the illness and nonspecific prodromal symptoms early in the course(5).

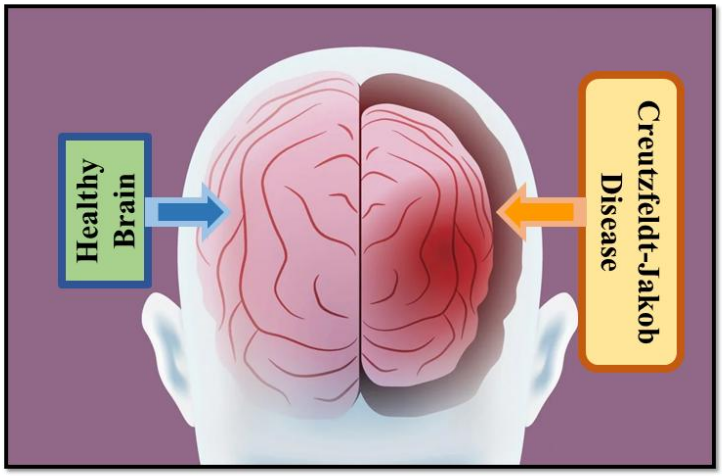

Figure 1: The Differences between a Healthy Brain and a Brain afflicted with Creutzfeldt-Jakob Disease (CJD)

EPIDEMIOLOGY

Globally, CJD affects roughly one person out of every million annually. In the United States, about 350 cases are diagnosed each year. The most prevalent type of prion disease in humans is sporadic CJD. Although it has been documented in both younger and older age groups, the condition's mean onset age is 62. The ratio of males to females in sporadic CJD is 1:1. Every year, there are about 1 to 2 new instances of sporadic CJD for every 1,000,000 people worldwide. Almost 70% of patients die within a year of the condition's beginning. Ninety per cent of individuals with sporadic CJD pass away within a year, with a mean lifespan of four to eight months.The second most prevalent form of CJD is genetic. Patients frequently have autosomal-dominant mutations in the PRNP gene with a family history. Less than 1% of instances are acquired, and they typically occur in young adults (mean age: 29).Between 1971 and 1990, 30 instances of Creutzfeldt-Jakob disease (CJD) were observed in India, 20 of which were confirmed and 10 of which were suspected. Similarities to previously published studies from other parts of the world have been found

through demographic research. Although two centres-Bombay and Bangalore—accounted for 21 (70%) of the cases, indicating clustering, this appears to be more apparent than actual. One participant was employed in the medical industry, where it was impossible to completely rule out the risk of iatrogenic transmission. There was no positive family history of CJD in any of the cases.Also referred to as subacute spongiform encephalopathy, Creutzfeldt Jakob Disease (CJD) is a neurological illness that progresses quickly and is fatal. Sporadic CJD, which accounts for 85% of known cases, genetic or familial CJD, which is defined by mutations in the human prion protein (PRNP) gene (10–15% of cases), and iatrogenic CJD, which is caused by unintentional transmission through medical and surgical procedures (1–2% of cases), are the three main subtypes of CJD. There is one case of CJD for every million people worldwide each year. There is a wealth of information on the incidence and prevalence of CJD in both Europe and America. Even though some Asian nations have their own CJD monitoring units, Asia is still not included in the global surveillance database.In Asia, sCJD is the most common type of CJD. Hong Kong (1) has reported the fewest cases of sCJD, while China (1957) and Japan (1705) have reported the most. Conversely, India has the lowest gCJD (2) while Japan has the highest (370)(6–10).

ETIOLOGY

A prion protein (PrP) is a typical neuron protein that is composed primarily of random coils and α-helices. Proteinase K-resistant β-pleated sheet aggregates make up the majority of proteinaceous infectious particles, commonly referred to as "prions," which are self-propagating proteins devoid of nucleic acid. When prions attach to normal PrP cellular isoforms, they transform α-helices into indigestible β-pleated sheets, which is how they replicate. CJD and other transmissible spongiform encephalopathies, such as kuru, scrapie, and bovine spongiform encephalopathy (mad cow disease), are brought on by these particles. The mode of transmission can be used to categorize CJD. The most prevalent kind of CJD (about 85%), sporadic, is caused by normal PrP isoform misfolding without any obvious triggers. Variably protease-sensitive prionopathy and sporadic fatal insomnia are subtypes of sporadic CJD. A heritable genetic mutation is the cause of genetic CJD, which is the second most prevalent kind (about 10–15%). Gerstmann-Sträussler-Scheinker syndrome, fatal familial insomnia, and familial CJD are subtypes of this illness.Less than 1% of cases have infectious CJD, which is caused by external prion transfer. Kuru, iatrogenic CJD, and variant CJD are subtypes of infectious CJD. Before the practice was outlawed in the 1950s, the Fore people of Papua New Guinea practised ceremonial cannibalism, which involved eating the brains of deceased family members. This practice is known as Kuru. Unintentional prion inoculation during operations results in iatrogenic CJD. By a process akin to that of bovine spongiform encephalopathy, variant CJD is linked to the consumption of contaminated beef. The majority of variant CJD cases that have been reported come from France and the United Kingdom.(11–13).

PATHOPHYSIOLOGY

Either naturally or as a consequence of PrPSc infection, normal cellular prion protein (PrPc) changes into the disease-causing form PrP scrapie (PrPSc). PrPSc spreads by itself and builds up all across the brain. Affected neurons exhibit abnormalities in intracellular protein folding, ubiquitination, and trafficking due to the very chemically stable β-pleated aggregates. Additionally, astrocytes may react to prion-induced harm by swelling and degrading. These alterations lead to neurodegeneration. Abnormalities may not be visible upon gross inspection of affected brains. Nonetheless, light microscopy frequently reveals the following characteristics (see image). Confirmation of Creutzfeldt-Jacob Disease by Neuropathology: Particularly in the cerebral cortex, caudate nucleus, thalamus, putamen, and molecular layer of the cerebellum, vacuolation or spongiform degeneration loss of neurons, fibrous proliferation of astrocytes, primarily in the grey matter, or astrocyte gliosis(14).

SIGNS AND SYMPTOMS

Changes in mental capacities are a hallmark of Creutzfeldt-Jakob disease. The symptoms typically worsen in a matter of weeks to months. Personality changes are among the early indications. Loss of memory. Cognitive impairment. Blindness or blurred vision. Sleeplessness. Coordination issues.Speakingisdifficult.Difficulty swallowing. Jerky, abrupt motions. Usually, death happens within a year. The majority of Creutzfeldt-Jakob disease patients pass away from related medical conditions. These could include pneumonia or other infections, heart problems, lung failure, falls, or difficulty swallowing.Changes in mental capacities may be more noticeable in the early stages of CJD in individuals with variant CJD. Dementia frequently appears later in the course of the illness. The inability to reason, think, or remember are all signs of dementia. Compared to CJD, variant CJD affects persons earlier in life. It seems that variant CJD lasts 12 to 14 months. Variably protease-sensitive prionopathy is another uncommon type of prion disease (VPSPr). It may resemble dementia in various ways. It results in cognitive changes as well as issues with thinking and speaking. The illness lasts for roughly 24 months, which is longer than other prion disorders.(15–23).

DIAGNOSIS

Because it manifests similarly to other illnesses that present with RPD, Creutzfeldt-Jakob disease is frequently difficult to diagnose. The suggested first screening tests for assessing RPD are as follows: Whole blood count. Completely metabolic panel. Magnesium in the blood. Rapid reagin of plasma. Rate of erythrocyte sedimentation. Antibodies against anti-nuclear. C-reactive protein. Testing for thyroid function. Level of vitamin B12. HIV test. Titer forLymedisease. Autoimmuneantibodies. Urine analysis. CSF tests, such as cell count with differential, oligoclonal bands, and glucose. The CSF 14-3-3 protein is a prion disease test. VDRL test (Venereal Disease Research Laboratory).The first imaging test that may be ordered is computed tomography (CT). Diffusion-weighted imaging (DWI) and fluid-attenuated inversion recovery (FLAIR), two related modalities, and brain magnetic resonance imaging (MRI) with or without contrast, however, can provide more information on the afflicted areas. When imaging is paired

with an electroencephalogram (EEG), clinical, and laboratory results, CJD can be identified.Based on the clinical, EEG, and CSF results, the World Health Organization released diagnostic criteria for CJD in 1998. However, those standards are no longer relevant due to sophisticated diagnostic techniques like MRI and genetic testing. Brain MRI was found to be accurate in around 90% of cases and is a more sensitive and specific test for variant CJD than CSF 14-3-3 protein (see Image). An early-stage Creutzfeldt-Jakob disease patient's brain MRI.In sporadic CJD, anomalies in the deep nuclei and cortical grey matter (cortical ribboning) are frequently seen by brain MRI using T2-weighted scanning, DWI, and apparent diffusion coefficient (ADC) sequences. The sensitivity and specificity of MRI with DWI or FLAIR imaging are 98% and 93%, respectively. Usually, DWI shows hyperintensities in the brain, thalamus, and basal ganglia. Although it can also be observed in other types of CJD, the "hockey stick" or "pulvinar" indication denotes variant, infectious, or acquired CJD. When paired with the usual EEG abnormalities, CSF 14-3-3 may be more sensitive for CJD than other prion disorders.Rapid neurodegeneration is indicated by CSF protein biomarkers such as the 14-3-3 protein, total tau (T-tau), and neuron-specific enolase (NSE). Although they are not specific to CJD, these tests can aid in its diagnosis. Although both tests can yield significant false-negative and false-positive findings, a high tau level (more than 1150 picogram/mL) offers greater accuracy and specificity than 14-3-3 protein as a diagnostic test for CJD.The American Academy of Neurology advised in 2012 to request CSF 14-3-3 only in cases when there is a high suspicion of CJD. Recently, DWI was reported to have a 97% diagnosis accuracy when compared to MRI using these non-prion-specific CSF biomarkers. This was greater than the T-tau (79.6%), 14-3-3 protein (70.4%), and NSE (71.4%) assays. About three-fourths of the time, these conventional surrogate marker proteins may be accurately detected. In patients with CJD, routine CSF analyses that measure oligoclonal IgG, total protein, white blood cell count, total cell count, and glucose are typically unremarkable.A new diagnostic test known as "second-generation Real Time-Quaking-Induced Conversion (RT-QuIC)," which is extremely sensitive and specific for CJD, was introduced by the National Prion Disease Pathology Surveillance Center in April 2015. In the CSF of individuals with CJD, RT-QuIC can reliably identify pathogenic prion proteins. Unlike the 14-3-3 protein, T-tau, and NSE, which are indirect tests, RT-QuIC directly identifies the pathogenic prion protein. According to a small number of investigations, RT-QuIC shows a high specificity (about 98%) and a reasonable sensitivity (around 80%) for sporadic CJD. CSF RT-QuIC tests are negative in the majority of patients with the VV2 sporadic CJD subtype. RT-QuIC is frequently positive in various types of hereditary prion disease, some of which do not exhibit the typical sporadic CJD MRI signs, although it is not as sensitive as the MRI.Using olfactory epithelium brushings instead of CSF as a specimen may increase RT-QuIC's sensitivity. According to recent research, the accuracy of RT-QuIC in diagnosing CJD is on par with brain biopsy. Brain biopsies are more invasive than RT-QulC. However, more recent, less intrusive diagnostic techniques are not covered by the current CJD standards. Periodic sharp wave complexes (PSWCs) are often found in the late stages of CJD, but EEG is not as sensitive as brain MRI or CSF investigations in identifying the disorder. Sixty-seven to ninety-five per cent of individuals with sporadic CJD have bi- or triphasic PSWC(24–28). The following characteristics define this discovery, which is regarded as supporting rather than diagnostic: complexities that are lateralized or generalized. To rule out semiperiodic activity, there must be a minimum of five repeating periods with a duration difference of less than 500 milliseconds. strictly periodic brain potentials, most of which have a duration of 100–600 ms and an inter-complex interval of 500–2000 ms.While the MV2 subtype seldom displays PSWCs, the MM1 and MV2 subtypes commonly do so in patients with sporadic CJD. Patients with kuru, variant CJD, Gerstmann-Sträussler-Scheinker syndrome, or fatal familial insomnia do not have PSWCs. Patients with genetic CJD, especially those with codon 200 mutations, can infrequently exhibit PSWCs. The diagnosis of CJD is confirmed by brain tissue biopsy or postmortem brain examination. But not every part of the brain is impacted by the illness. Since aberrant characteristics are most likely to be discovered in subcortical regions, imaging examinations must focus on these areas. In addition to being harmful, surgery might not always remove the damaged brain tissue. The clinical fate of the patient remains unchanged upon diagnosis of CJD.Vascular, neurodegenerative, autoimmune, infectious, thromboembolic, neoplastic, iatrogenic, and toxic metabolic disorders are all included in the wide differential diagnosis of RPD. RPD can also result from vascular disorders such as hypertensive encephalopathy, cerebral myeloid angioplasty, multiple infarcts, or stroke. RPD can potentially be a symptom of intravascular lymphoma or vasculitis. A comprehensive medical evaluation may be necessary to differentiate these disorders from CJD. Imaging tests can assist in ruling out neoplastic and vascular causes. Autoimmune, viral, metabolic, and neurodegenerative diseases can all be identified by blood and CSF tests.Brain biopsies are therefore only recommended in cases when a reversible disorder is considered in the differential diagnosis. Patients with symptomatic variant CJD have been found to contain prions in their blood and urine. Until prion infectivity has been ruled out, care should be taken while handling bodily fluids and tissues from patients with variant CJD.(29).

DISEASE WITH CREUTZFELDT-JAKOB SOME TECHNIQUES TOWARDS BENEFICIAL MANAGEMENT AND TREATMENT

Since there is no known cure for CJD, supportive care is the cornerstone of treatment. To present, the majority of CJD experimental medications have not shown any discernible advantages. However, investigations on rodents have demonstrated that intraventricular pentosan polysulfate inhibits the development of PrPSc. Four patients showed an apparent prolongation in life from 37 to 114 months. To develop a treatment for this deadly illness, more investigation is required. Families at risk for the hereditary form of CJD may benefit from early discovery of a PRNP mutation. Individuals with the illness can plan for end-of-life issues. Patients' quality of life may be enhanced by supportive treatment and psychosocial support. Family planning and genetic counselling can help stop the spread of disease to children

born to people with PRNP mutations. To provide the best treatment possible to those impacted by CJD, team members must work together. The majority of CJD patients pass away within a year after their symptoms appear. Hospice care arrangements need to be made right away. Since there is no effective treatment for the illness and death is certain, family counselling is crucial. When combined with family counselling and the right palliative care, interprofessional collaboration can help slow the disease's deadly progression.(30).

ISSUES WITH CREUTZFELDT-JAKOB DISEASE

Complications of CJD include both psychological and physical issues. CJD sufferers frequently distance themselves from friends and family, eventually losing the capacity to identify or connect with them. Additionally, patients lose the ability to take care of themselves and frequently end up in a coma. The fatality rate for CJD is 100%.Despite all the advancements that have helped us comprehend CJD, the outlook is very dismal. The illness is always lethal. Except in certain situations, death happens within a year of the commencement of symptoms(31,32).

CONCLUSION AND FUTURE DIRECTION

An overview of Creutzfeldt-Jakob disease, including its several causes, epidemiology, pathophysiology, and prevention, opens our review articles. Our research indicates that Creutzfeldt-Jakob disease cannot be helped by drugs or other therapies. Only psychosocial help and supportive care. More randomized controlled research is required to address Creutzfeldt-Jakob disease. We would like to conduct more studies on Creutzfeldt-Jakob disease in the future. Future counseling-based research in our state or nation will be able to evaluate patients' physical and mental health and provide more precise information on Creutzfeldt-Jakob disease and its treatment thanks to the collaboration of our colleagues.

ETHICAL STATEMENT

Be sincere and maintain a high standard of conduct in all of our dealings and activities about our jobs. Let us be honest in our words and actions.

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CONFLICT OF INTEREST

The authors attest that they are free of any known financial or personal conflicts of interest that would taint the findings of this study.

INFORMED CONSENT

Using websites, review articles, and other sources to produce research content.

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