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Research Article

CJD CREUTZFELDT-JAKOB DISEASE - (PRION)

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A R T I C L E I N F O

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A B S T R A C T

Creutzfeldt - Jakob disease (CJD) progressively destroys brain cells, and it causes tiny holes in the brain. People with CJD will have ataxia, or difficulty controlling body movements, abnormal gait, speech, and dementia. Prion infections also cause small holes to develop in the brain, so it becomes sponge-like. The damage to the brain causes the mental and physical impairment associated with CJD and eventually leads to death. Sporadic CJD is more likely to occur in people who have specific versions of the prion protein gene. Familial or inherited CJD is a rare form of CJD. **Electroencephalogram (EEG)** measures the brain's patterns of electrical activity similar to the way an electrocardiogram (ECG) measures the heart's electrical activity.CJD caused by an inherited mutation (abnormality) in the gene that produces the prion protein. Opiate drugs may be prescribed to patients with CJD to help relieve pain, while clonazepam and sodium evaporate may help relieve involuntary muscle jerks.

INTRODUCTION

Prion diseases also referred to as the subacute spongiform encephalopathies. These diseases have a subacute to chronic clinical course with a similar neuropathology. All the diseases are transmissible and induced by an abnormal misfolded form of the prion protein that is extremely resistant to physical and chemical inactivation. The unusual nature of the transmissible agent and the emergence of variant Creutzfeldt-Jakob disease (as a result of ingestion of contaminated beef) have had a significant impact on public health in addition to science and medicine. New diagnostic tests, protein misfolding cyclic amplification, real time quaking-induced conversion, and new ideas about treatment of the subacute spongiform encephalopathies

Creutzfeldt-Jakob disease is a rare neurodegenerative disease that rapidly, progressively and severely affects the brain

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Creutzfeldt-Jakob disease (CJD) is caused by an abnormal infectious protein in the brain called a prion

Proteins are molecules, made up of amino acids, which help the cells in our body to function. They begin as a string of amino acids that then fold themselves into a three-dimensional shape. This 'protein folding' allows them to perform useful functions within our cells.

Normal (harmless) prion proteins are found in almost all body tissues, but at highest levels in brain and nerve cells.

The exact role of the normal prion proteins is unknown, but it's thought they may play a role in transporting messages between certain brain cells.

Mistakes sometimes occur during protein folding and the prion protein can't be used by the body. Normally, these misfolded prion proteins are recycled by the body, but if they're not they can build up in the brain.

How prions cause CJD

Prions are misfolded prion proteins that build up in the brain and cause other prion proteins to misfold as well. This causes the brain cells to die, releasing more prions to infect other brain cells.

Eventually, clusters of brain cells are killed and deposits of misfolded prion protein, called plaques, may appear in the brain.

Prion infections also cause small holes to develop in the brain, so it becomes sponge-like. The damage to the brain causes the mental and physical impairment associated with CJD and eventually leads to death.

Prions can survive in nerve tissue, such as the brain or spinal cord, for a very long time, even after death.

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Types of CJD

The different types of CJD are all caused by a build-up of prions in the brain. However, the reason why this happens is different for each type.

The causes of the main types of CJD are described below.

Sporadic CJD

Even though sporadic CJD is very rare, it's the most common type of CJD, accounting for around 8 in every 10 cases.

It's not known what triggers sporadic CJD, but it may be that a normal prion protein spontaneously changes into a prion, or a normal gene spontaneously changes into a faulty gene that produces prions.

Sporadic CJD is more likely to occur in people who have specific versions of the prion protein gene. At present, nothing else has been identified that increases your risk of developing sporadic CJD.

Variant CJD

In 2000, a government inquiry concluded that the prion was spread through cattle that were fed meat-and-bone mix containing traces of infected brains or spinal cords. The prion then ended up in processed meat products, such as beef burgers, and entered the human food chain.

Strict controls have been in place since 1996 to prevent BSE entering the human food chain and the use of meat-and-bone mix has since been outlawed.

It appears that not everyone who is exposed to BSE-infected meat will go on to develop CJD.

All definite cases of CJD occurred in people with a specific version (MM) of the prion protein gene, which affects how the body makes a number of amino acids. It's estimated that up to 40% of the UK population have this version of the gene.

Cases of vCJD peaked in the year 2000, in which there were 28 deaths from this type of CJD. There were no confirmed deaths in 2014. Some experts believe that the food controls have worked and that further cases of vCJD will continue to decline, but this doesn't rule out the possibility that other cases may be identified in future.

It's also possible for vCJD to be transmitted by blood transfusion, although this is very rare and measures have been put in place to reduce the risk of it happening.

We don't know how many people in the UK population could develop vCJD in the future and how long it will take for symptoms to appear, if they ever will.

A study published in October 2013 that involved testing random tissue samples suggested that around 1 in 2,000 people in the UK population may be infected with vCJD, but show no symptoms to date.

Familial or inherited CJD

Familial or inherited CJD is a rare form of CJD caused by an inherited mutation (abnormality) in the gene that produces the prion protein. The altered gene seems to produce misfolded prions that cause CJD.

Everyone has two copies of the prion protein gene, but the mutated gene is dominant. This means you only need to inherit one mutated gene to develop the condition. Therefore, if one of the parents has the mutated gene, there's a 50% chance it will be passed on to their children.

As the symptoms of familial CJD don't usually begin until a person is in their 50s, many people with the condition are unaware that their children are also at risk of inheriting this condition when they decide to start a family.

Iatrogenic CJD

Iatrogenic CJD (iCJD) is where the infection is spread from someone with CJD through medical or surgical treatment.

Most cases of iatrogenic CJD have occurred through the use of human growth hormone, which is used to treat children with restricted growth. Between 1958 and 1985, thousands of children were treated with the hormone, which at the time was extracted from the pituitary glands (a gland at the base of the skull) of human corpses.

A minority of those children developed CJD, as the hormones they received were taken from glands infected with CJD. Since 1985, all human growth hormone in the UK has been artificially manufactured, so there's now no risk. However, a small number of patients exposed before 1985 are still developing iCJD.

A few other cases of ICED have occurred after people received transplants of infected dura (tissue that covers the brain) or came into contact with surgical instruments that were contaminated with CJD. This happened because prions are tougher than viruses or bacteria, so the normal process of sterilising surgical instruments had no effect.

Once the risk was recognised, the Department of Health tightened the guidelines on organ donation and the reuse of surgical equipment. As a result, cases of iCJD are now very rare.

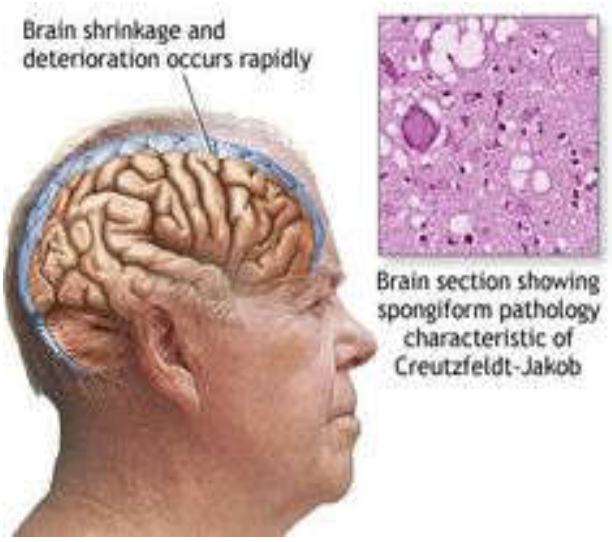
Pathophysiology

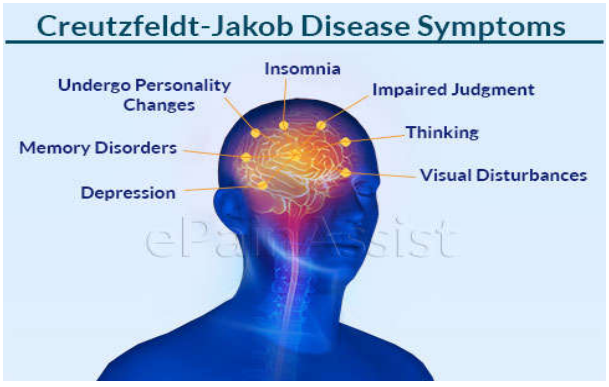
The transformation of prion proteins into prions and/or the resulting accumulation of prions in the CNS causes nerve cell injury and eventually death. During the process of nerve cell injury, fluid-filled vesicles appear in the dendritic tree of neurons. These vesicles are called vacuoles and enlarge to 10 to 20 micrometres or larger and cause the brain tissue to have a spongiform appearance when examined under the microscope, hence the old name for prion diseases, transmissible spongiform encephalopathies. This vacuolation (or spongiform) change is a hallmark characteristic of this disease, although other conditions can also have these microscopic changes. Other microscopic features of Creutzfeldt-Jakob disease (CJD) brain tissue include neuron loss, astrogliosis, and, most importantly, the presence of PrPSc by immunohistochemistry or western blot. In approximately 10% of CJD, amyloid plaques of PrPSc are found; and in cases of Gerstmann-Straussler-Scheinker, an amyloid PrPSc core is surrounded by another group of smaller amyloid globules. vCJD also has a relatively unique pathological feature of core PrPSc amyloid plaques surrounded by vacuoles; these are called florid plaques, as they are thought to resemble a flower. PrPSc plaques and pericellular deposits are found throughout the cerebrum and cerebellum in vCJD. In vCJD, PrPSc can also be identified in

the lymphoreticular system during the disease course; hence, tonsil biopsies often show PrPSc in vCJD, but not in other forms of human prion disease. At the cellular level, although much work has been done, we still do not understand the processes that lead to neuronal injury and death in prion disease, although oxidative stress may play an important role. There appears to also be damage to the GABAergic system in prion disease. One issue that is clear is that the prion protein, PrPC, is required, and may be sufficient, for development of prion disease

Symptoms

Specific Creutzfeldt-Jakob disease symptoms experienced by an individual:





- Depression
- Agitation, apathy and mood swings
- Rapidly worsening confusion, disorientation, and problems with memory, thinking, planning and judgment
- Difficulty walking
- Muscle stiffness, twitches and involuntary jerky movements

Diagnosis

Rapid symptom progression is one of the most important clues that a person may have Creutzfeldt-Jakob disease.

There is no single test-or any combination of tests-that can conclusively diagnose sporadic CJD in a living person, but the following tests may help determine whether an individual has CJD:

- Electroencephalogram (EEG) measures the brain's patterns of electrical activity similar to the way an electrocardiogram (ECG) measures the heart's electrical activity.
- Brain magnetic resonance imaging (MRI) can detect certain brain changes consistent with CJD.
- Lumbar puncture (spinal tap**)** tests spinal fluid for the presence of certain proteins.

Treating symptoms of CJD

For more information about how some of the specific symptoms of CJD may be treated see:

- treating ataxia (loss of physical co-ordination)
- treating urinary incontinence (loss of bladder control)
- treating bowel incontinence (loss of bowel control)
- treating dysphagia (swallowing difficulties)
- treating dystonia (muscle spasms and stiffness)
- help and support for blindness or vision loss

Care and support in the advanced stages of CJD

As CJD progresses, people with the condition will need significant nursing care and practical support.

As well as help with feeding, washing and mobility, some people may also need help urinating. A catheter (a tube that's inserted into the bladder and used to drain urine) is often required.

Many people will also have problems swallowing, so they may have to be given nutrition and fluids through a feeding tube.

It may be possible to treat people with CJD at home, depending on the progression and severity of the condition.

Caring for someone with CJD can be distressing and difficult to cope with, so many carers prefer to use the specialist services of a hospital or hospice.

A number of drugs have been tested to treat the disease, including steroids, antibiotics and antiviral medicines, however, treatment of the disease generally revolves around alleviating pain and other uncomfortable symptoms.

Opiate drugs may be prescribed to patients with CJD to help relieve pain, while clonazepam and sodium valproate may help relieve involuntary muscle jerks

Acyclovir, amantadine, antibiotics, anti-viral agents, interferon and steroids have all been tried as potential therapies for patients with CJD

Complications

Most patients lapse into a coma as the disease progresses and the symptoms worsen. The cause of death is usually due to heart failure, respiratory failure, pneumonia or other infections.

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