CREUTZFELDT-JACOB DIS pdf

1: Creutzfeldt-Jakob Disease, Classic (CJD) | Prion Diseases | CDC

Creutzfeldt-Jakob disease (CJD) is a rare, degenerative, fatal brain disorder. It affects about one person in every one million per year worldwide; in the United States there are about cases per year.

CJD occurs throughout the world at an incidence of one in every one million people. Among certain populations, such as Libyan Jews, rates are somewhat higher. CJD is similar to other neurodegenerative diseases such as kuru , a human disorder, and scrapie , which occurs in sheep and goats. All three diseases are types of transmissible spongiform encephalopathies , so called because of the characteristic spongelike pattern of neuronal destruction that leaves brain tissue filled with holes. Causes and symptoms CJD, as well as other spongiform encephalopathies, is caused by an unusual pathogenic agent called a prion. A prion is a deviant form of a normally harmless protein found in the brains of mammals and birds. As prions replicateâ€"by converting normal forms of the protein into their abnormal shapeâ€"they accumulate within nerve cells, causing neurodegeneration. CJD commonly occurs in adults between ages 40 and 70, although some young adults have been stricken with the disease. Both men and women are affected equally. The onset of the disease is usually characterized by vague psychiatric or behavioral changes, which are followed within weeks or months by a progressive dementia that is often accompanied by abnormal vision and involuntary movements. The disease is usually fatal within a year of symptom onset. Types There are three major types of CJD: In these cases it is unclear what molecular process causes the prion protein to appear in the first place. Between 5 and 15 percent of CJD cases show a familial pattern of inheritance. In these inherited cases a mutation in a gene designated PRNP , which encodes the prion protein PrP, is passed from parent to child in a dominant fashion i. More than 50 different mutations in PRNP have been identified. In addition, mutations have been identified that do not cause disease but may render individuals more susceptible to infection with the prion. These latter mutations may be involved in some of the sporadic incidences of the disease. Transmission There is no evidence that a person with CJD is contagious. The rare cases of the disease that arise from human-to-human transmission are considered forms of iCJD essentially physician-induced CJD , having been caused by exposure to the prion during medical procedures. Such accidental transmission has occurred in corneal transplants, through the use of contaminated medical or surgical instruments, and through the transfusion of contaminated blood products, including prion-infected plasma. Transmission also may have occurred through the injection of growth hormone derived from human pituitary glands. Although human-to-animal prion transmission has been demonstrated in the laboratory, researchers are not sure whether prions that cause disease in one species can give rise to a prion disease in humans. There is increasing evidence that these cases resulted from the consumption of tissues notably nerve tissue contaminated with the prion that causes bovine spongiform encephalopathy BSE , or mad cow disease. Chronic wasting disease, which is caused by a prion that occurs in elk and deer, is similar in nature to BSE. Although there have been no cases of animal-to-human transmission of this prion, researchers suspect that eating contaminated tissues of deer and elk could give rise to another variant form of CJD. As a result, scientists have been monitoring cases of CJD in areas where chronic wasting disease is endemic. Diagnosis and treatment Diagnosis of CJD typically entails spinal tap , electroencephalography , and other procedures to assess neurological function in order to rule out conditions that might produce similar symptoms. Diagnosis is confirmed through brain biopsy , in which a small section of tissue is removed from the brain and examined in a laboratory. Scientists are developing tests capable of detecting prions in cerebrospinal fluid and blood. Such tests could enable early diagnosis and improve prion screening for blood transfusions. There is no known cure for CJD, nor can the progression of the disease be delayed by medication or surgery. Hence, treatment is supportive, being aimed primarily at minimizing pain and discomfort. Learn More in these related Britannica articles:

CREUTZFELDT-JACOB DIS pdf

2: Creutzfeldt-Jakob disease - Diagnosis and treatment - Mayo Clinic

Creutzfeldt-Jakob (KROITS-felt YAH-kobe) disease is a degenerative brain disorder that leads to dementia and, ultimately, death. Symptoms of Creutzfeldt-Jakob disease (CJD) can resemble those of other dementia-like brain disorders, such as Alzheimer's.

In most people with CJD, these symptoms are accompanied by involuntary movements and the appearance of an atypical, diagnostic electroencephalogram tracing. The duration of the disease varies greatly, but sporadic non-inherited CJD can be fatal within months or even weeks. When brain tissue from a person with CJD is examined under a microscope , many tiny holes can be seen where whole areas of nerve cells have died. The word "spongiform" in " transmissible spongiform encephalopathies " refers to the sponge-like appearance of the brain tissue. Cause[edit] Transmissible spongiform encephalopathy diseases are caused by prions. Prions are proteins that occur normally in neurons of the central nervous system CNS. These proteins, once misfolded, are thought to affect signaling processes, damaging neurons and resulting in degeneration that causes the spongiform appearance in the affected brain. This mass of misfolded proteins disrupts neuronal cell function and causes cell death. Mutations in the gene for the prion protein can cause a misfolding of the dominantly alpha helical regions into beta pleated sheets. This change in conformation disables the ability of the protein to undergo digestion. Once the prion is transmitted, the defective proteins invade the brain and induce other prion protein molecules to misfold in a self-sustaining feedback loop. These neurodegenerative diseases are commonly called prion diseases. In sporadic cases the misfolding of the prion protein probably occurs as a natural, spontaneous process. All types of CJD are transmissible irrespective of how they occur in the patient. The World Health Organization and the US Centers for Disease Control and Prevention recommend that instrumentation used in such cases be immediately destroyed after use; short of destruction, it is recommended that heat and chemical decontamination be used in combination to process instruments that come in contact with high-infectivity tissues. No cases of iatrogenic transmission of CJD have been reported subsequent to the adoption of current sterilization procedures, or since Electroencephalography â€” may have characteristic generalized periodic sharp wave pattern. Periodic sharp wave complexes develop in half of the patients with sporadic CJD, particularly in the later stages. However, a positive result should not be regarded as sufficient for the diagnosis. MRI of the brain â€” often shows high signal intensity in the caudate nucleus and putamen bilaterally on T2-weighted images. In recent years, studies have shown that the tumour marker Neuron-specific enolase NSE is often elevated in CJD cases; however, its diagnostic utility is seen primarily when combined with a test for the protein. Imaging findings are variable in their appearance, and also variable in sensitivity and specificity. Of the MRI sequences, diffuse-weighted imaging sequences are most sensitive. Characteristic findings are as follows: Due to its invasiveness, biopsy will not be done if clinical suspicion is sufficiently high or low. A negative biopsy does not rule out CJD, since it may predominate in a specific part of the brain. Neuronal loss and gliosis are also seen. However, extra-neuronal vacuolization can also be seen in other disease states. These vacuoles appear clear and punched-out. Larger vacuoles encircling neurons, vessels, and glia are a possible processing artifact. Medical procedures that are associated with the spread of this form of CJD include blood transfusion from the infected person, use of human-derived pituitary growth hormones, gonadotropin hormone therapy, and corneal and meningeal transplants.

CREUTZFELDT-JACOB DIS pdf

3: Creutzfeldt-Jakob Disease | CJD | MedlinePlus

Five New Zealanders have been confirmed to have died of the sporadic form of Creutzfeldt-Jakob disease (CJD) in United States. In , there was a confirmed death from CJD of a person from Manchester, New Hampshire.

Print Diagnosis Only a brain biopsy or an examination of brain tissue after death autopsy can confirm the presence of Creutzfeldt-Jakob disease. But doctors often can make an accurate diagnosis based on your medical and personal history, a neurological exam, and certain diagnostic tests. The exam is likely to reveal characteristic symptoms such as muscle twitching and spasms, abnormal reflexes, and coordination problems. People with CJD also may have areas of blindness and changes in visual-spatial perception. In addition, doctors commonly use these tests to help detect CJD: This imaging technique uses radio waves and a magnetic field to create cross-sectional images of your head and body. Cerebral spinal fluid surrounds and cushions your brain and spinal cord. In a test called a lumbar puncture â€" popularly known as a spinal tap â€" doctors use a needle to withdraw a small amount of this fluid for testing. Treatment No effective treatment exists for Creutzfeldt-Jakob disease or any of its variants. A number of drugs have been tested and have not shown benefits. For that reason, doctors focus on alleviating pain and other symptoms and on making people with these diseases as comfortable as possible. In some cases when you call for an appointment, you may be referred immediately to a brain specialist neurologist. What you can do List your symptoms, including any that may seem unrelated to the reason for which you scheduled the appointment. Write down key personal information, including recent life changes. List medications, vitamins and supplements you take. Bring a family member or friend along, if possible. Someone who accompanies you might help you remember something you missed or forgot. Write down questions to ask your doctor. For Creutzfeldt-Jakob disease, some basic questions to ask your doctor include: What is likely causing my symptoms? Other than the most likely cause, what are other possible causes for my symptoms? What tests do I need? What is the best course of action? Are there restrictions I need to follow? Should I see a specialist? I have other medical conditions. How do I manage them together? Are there brochures or other printed material I can have? What websites do you recommend? What to expect from your doctor Your doctor is likely to ask you a number of questions, including: When did your symptoms begin? Have your symptoms been continuous or occasional? How severe are your symptoms? What, if anything, seems to improve your symptoms? What, if anything, appears to worsen your symptoms? Has anyone in your family had Creutzfeldt-Jakob disease? Have you lived or traveled extensively outside the United States?

CREUTZFELDT-JACOB DIS pdf

4: Creutzfeldtâ€“Jakob disease - Simple English Wikipedia, the free encyclopedia

Creutzfeldt-Jakob disease is a very rare disorder that causes the brain to break down. Also called "classic" CJD, it worsens quickly. Most people die within a year of getting it.

What Causes Creutzfeldt-Jakob Disease? CJD is caused by an infectious agent called a prion. Prions are a type of small protein that are normally found in the tissues of many mammals. In prion disease, these proteins are abnormally folded, and form clumps. When they infect a mammal, its normal proteins start to take on the incorrect structure of the infectious prions. This causes brain injury by destroying nerve cells and disrupting the structure of your brain. On brain imaging, people with CJD appear to have holes in their brains where cells have died â€" causing their brains to resemble a sponge. However, it most commonly affects people in their late 50s. Sporadic CJD has no connection to mad cow disease. Sporadic CJD occurs when normal proteins spontaneously mutate to the abnormal prion type. Sporadic CJD is most common in people over the age of It occurs when you inherit a mutated gene associated with prion disease from a parent. People with inherited CJD often have family members with the disease. The extent of how CJD manifests in separate family members can vary widely and is known as variable expressivity. However, your risk of eating infected meat is very low. You can also become infected after receiving blood or transplanted tissues, such as a corne, from an infected donor. Fortunately, there is rigorous sterilization protocols for instruments that have been in contact with tissue at risk for prion exposure, such as brain or eye tissue. Despite all the press on mad cow disease, vCJD is very rare. Less than 1 percent of people with CJD have the variant type. The risk of classic CJD increases with age. Instead, you need to be exposed to infected bodily fluids or tissue. Caregivers of people with CJD should take extra precautions to lower their risk of contracting the disease: Protect your hands and face from exposure to body fluids. Make sure to wash your hands, face, and all exposed skin before smoking, eating, or drinking. Use waterproof bandages to cover cuts or bruises. To diagnose CJD, your doctor will begin with a complete medical history, physical examination, and neurological evaluation. The rapid progression of symptoms distinguishes CJD from other causes of dementia. Your doctor can also use a number of tests to establish your diagnosis: It can detect small changes to your brain that may suggest CJD. An MRI uses magnetic fields to create images of your brain. Usually, the CAT scans will be normal. In some patients, rapid degeneration of brain tissue can be detected. Lumbar Puncture Single-Use Kit In this test, your doctor will use a thin needle to puncture the lining of your spinal cord to obtain spinal fluid. If your spinal fluid tests positive for elevated levels of a protein called , you may have CJD. However, high levels of protein are also found in many other diseases. EEG In this test, your doctor will use scalp electrodes to examine your brain waves. If you have CJD, your brain waves may show sharp spikes. Blood Tests Your doctor can use blood tests to identify and rule out problems such hypothyroidism and syphilis, which can also cause dementia. It is important to remember that only a biopsy of brain tissue can confirm a diagnosis of CJD. There is no known cure or effective treatment for CJD. However, medications can be used to treat some of the mental changes and personality abnormalities that occur. Your course of treatment will probably focus on making you comfortable and helping you function safely in your environment. What Is the Long-Term Outlook? People with variant CJD tend to survive a little longer, from the onset of symptoms to death. Symptoms of CJD will get worse until you lapse into a coma. The most frequent causes of death for people with CJD are:

CREUTZFELDT-JACOB DIS pdf

5: Variant Creutzfeldtâ€“Jakob disease - Wikipedia

Creutzfeldt-Jakob disease (CJD) is a rare, degenerative brain www.enganchecubano.comms usually start around age Memory problems, behavior changes, vision problems, and poor muscle coordination progress quickly to dementia, coma, and death.

Difficulty standing or walking How is CJD diagnosed? A biopsy is a procedure to remove small piece of tissue from your brain. Healthcare providers will send the sample to a lab to check for abnormal prions. An EEG, or electroencephalogram, is done to see activity in your brain. Healthcare providers will place small pads on your head and connect them to a machine to see how your brain is working. A lumbar puncture, or spinal tap, is a procedure during which healthcare providers insert a needle and remove fluid from around your spinal cord. It may show signs of inflammation or infection. A MRI may show areas of your brain that are damaged. You may be given contrast dye to help your brain show up better in the pictures. Tell the healthcare provider if you have ever had an allergic reaction to contrast dye. Do not enter the MRI room with anything metal. Metal can cause serious injury. Tell the healthcare provider if you have any metal in or on your body. How is CJD treated? There is no cure for CJD. The goal of treatment is to help reduce your symptoms. Medicines are given to decrease pain, anxiety, and muscle spasms. You may need treatment or therapy to help with speaking or swallowing. When should I contact my healthcare provider? You have a fever. You are depressed and feel you cannot cope with your condition. You are anxious or nervous even after you take medicine. You have severe muscle twitching or pain even after you take medicine. You have questions or concerns about your condition or care. When should I seek immediate care or call ? You have trouble breathing. You have sudden changes in your vision. Care Agreement You have the right to help plan your care. Learn about your health condition and how it may be treated. Discuss treatment options with your healthcare providers to decide what care you want to receive. You always have the right to refuse treatment. The above information is an educational aid only. It is not intended as medical advice for individual conditions or treatments. Talk to your doctor, nurse or pharmacist before following any medical regimen to see if it is safe and effective for you.

6: Creutzfeldt-Jakob Disease Fact Sheet | National Institute of Neurological Disorders and Stroke

About Creutzfeldt-Jakob disease Prion diseases, such as Creutzfeldt-Jakob disease, occur when prion protein, which is found throughout the body but whose normal function isn't yet known, begins folding into an abnormal three-dimensional shape.

How prions fold How prions fold Prions are proteins that occur naturally in the brains of animals and people. Creutzfeldt-Jakob disease and its variants belong to a broad group of human and animal diseases known as transmissible spongiform encephalopathies TSEs. The name derives from the spongy holes, visible under a microscope, that develop in affected brain tissue. The cause of Creutzfeldt-Jakob disease and other TSEs appears to be abnormal versions of a kind of protein called a prion. Normally these proteins are harmless. The three ways it develops are: Most people with classic CJD develop the disease for no apparent reason. Fewer than 15 percent of people with CJD have a family history of the disease or test positive for a genetic mutation associated with CJD. This type is referred to as familial CJD. A small number of people have developed CJD after being exposed to infected human tissue during a medical procedure, such as a cornea or skin transplant. Also, because standard sterilization methods do not destroy abnormal prions, a few people have developed CJD after undergoing brain surgery with contaminated instruments. Variant CJD is linked primarily to eating beef infected with mad cow disease bovine spongiform encephalopathy, or BSE. Risk factors Autosomal dominant inheritance pattern Autosomal dominant inheritance pattern In an autosomal dominant disorder, the mutated gene is a dominant gene located on one of the nonsex chromosomes autosomes. You need only one mutated gene to be affected by this type of disorder. A person with an autosomal dominant disorder â€” in this case, the father â€” has a 50 percent chance of having an affected child with one mutated gene dominant gene and a 50 percent chance of having an unaffected child with two normal genes recessive genes. Most cases of Creutzfeldt-Jakob disease occur for unknown reasons, and no risk factors can be identified. However, a few factors seem to be associated with different kinds of CJD. Sporadic CJD tends to develop later in life, usually around age Onset of familial CJD occurs slightly earlier and vCJD has affected people at a much younger age, usually in their late 20s. People with familial CJD have a genetic mutation that causes the disease. The disease is inherited in an autosomal dominant fashion, which means you need to inherit only one copy of the mutated gene, from either parent, to develop the disease. If you have the mutation, the chance of passing it on to your children is 50 percent. Exposure to contaminated tissue. The risk of contracting vCJD from eating contaminated beef is difficult to determine. In general, if countries are effectively implementing public health measures, the risk is virtually nonexistent. Complications As with other causes of dementia, Creutzfeldt-Jakob disease profoundly affects the brain as well as the body, although CJD and its variants usually progress much more rapidly. People with CJD usually withdraw from friends and family and eventually lose the ability to recognize or relate to them. They also lose the ability to care for themselves and many eventually slip into a coma. The disease ultimately is fatal. Prevention There is no known way to prevent sporadic CJD. If you have a family history of neurological disease, you may benefit from talking with a genetics counselor, who can help you sort through the risks associated with your situation. These measures have included: This includes people who: Have a biological relative who has been diagnosed with CJD Have received a dura mater brain graft Have received human growth hormone Spent at least three months in the United Kingdom from to Spent five years or more in Europe since Have lived at United States military bases located in Northern Europe for at least six months from to , or in other locations in Europe from to Received a blood transfusion in the U. Only three cases have been reported in the U. According to the Centers for Disease Control and Prevention, strong evidence suggests that these cases were acquired abroad â€” two in the United Kingdom and one in Saudi Arabia. In the United Kingdom, where the majority of vCJD cases have occurred, fewer than cases have been reported. CJD incidence peaked between and and has been declining since. A very small number of other vCJD cases have been reported in other countries worldwide. Regulating potential sources of vCJD Most countries have taken steps to prevent BSE-infected tissue from entering the food supply, including: Tight restrictions on importation of cattle from countries where BSE is common Restrictions on animal feed Strict

procedures for dealing with sick animals Surveillance and testing methods for tracking cattle health Restrictions on which parts of cattle can be processed for food.

CREUTZFELDT-JACOB DIS pdf

7: Creutzfeldt-Jakob disease: MedlinePlus Medical Encyclopedia

Creutzfeldt-Jakob disease (CJD) is a rare fatal brain disorder that usually occurs later in life and runs a rapid course. In the early stages of the disease, patients may have failing memory, behavior changes, impaired coordination, and vision problems.

MRI of the brain Spinal tap to test for a protein called The disease can only be confirmed with a brain biopsy or autopsy. Today, it is very rare for a brain biopsy to be done to look for this disease. Treatment There is no known cure for this condition. Different medicines have been tried to slow the disease. These include antibiotics, drugs for epilepsy, blood thinners, antidepressants, and interferon. But none works well. This may require monitoring and assistance in the home or in a care facility. Family counseling may help the family cope with the changes needed for home care. People with this condition may need help controlling unacceptable or dangerous behaviors. This involves rewarding positive behaviors and ignoring negative behaviors when it is safe. They may also need help getting oriented to their surroundings. Sometimes, medicines are needed to help control aggression. Persons with CJD and their family may need to seek legal advice early in the course of the disorder. Advance directive , power of attorney, and other legal actions can make it easier to make decisions about the care of the person with CJD. People with sporadic CJD are unable to care for themselves within 6 months or less after symptoms begin. The disorder is fatal in a short time, usually within 8 months. People who have variant CJD get worse more slowly, but the condition is still fatal. A few people survive for as long as 1 or 2 years. The cause of death is usually infection, heart failure, or respiratory failure. The course of CJD is: Infection with the disease Loss of ability to interact with others Loss of ability to function or care for oneself Death When to Contact a Medical Professional CJD is not a medical emergency. However, getting diagnosed and treated early may make the symptoms easier to control, give patients time to make advance directives and prepare for the end of life, and give families extra time to come to terms with the condition. Prevention Medical equipment that may be contaminated should be removed from service and disposed of. People known to have CJD should not donate a cornea or other body tissue. Most countries now have strict guidelines for managing infected cows to avoid transmitting CJD to humans. Prions and prion diseases of the central nervous system transmissible neurodegenerative diseases. Review provided by VeriMed Healthcare Network.

8: Creutzfeldt-Jakob disease | pathology | www.enganchecubano.com

Creutzfeldt-Jakob disease (CJD) is a rapidly progressive, invariably fatal neurodegenerative disorder believed to be caused by an abnormal isoform of a cellular glycoprotein known as the prion protein.

Blood products[edit] As of evidence suggest that while there may be prions in the blood of individuals with vCJD, this is not the case in individuals with sporadic CJD. A blood test for vCJD infection is possible [10] but is not yet available for screening blood donations. Significant restrictions exist to protect the blood supply. The UK government banned anyone who had received a blood transfusion since January from donating blood. Given the large number of U. Later changes to this policy have relaxed the restriction to a cumulative total of five years or more of civilian travel in European countries six months or more if military. The three-month restriction on travel to the UK, however, has not been changed. In , the NZBS further extended restrictions to permanently preclude donors having received a blood transfusion in the United Kingdom since January , and in April , restrictions were further extended to include the Republic of Ireland and France. They are also ineligible if they have spent a cumulative total of five years or more in Western Europe outside the U. Despite this the scientific consensus is that the risk is negligible, as there is no evidence Creutzfeldtâ€"Jakob is sexually transmitted. One explanation for this can be found in the genetics of patients with the disease. The human PRNP protein which is subverted in prion disease can occur with either methionine or valine at amino acid , without any apparent difference in normal function. Only a single person with vCJD tested was found to be heterozygous; most of those affected had two copies of the methionine-containing form. It is not yet known whether those unaffected are actually immune or only have a longer incubation period until symptoms appear. In the s, cannibalism was banned in Papua New Guinea. A critique to this theory is that while mortuary cannibalism was banned in Papua New Guinea in the s, that does not necessarily mean that the practice ended. Fifteen years later Jared Diamond was informed by Papuans that the practice continued. These researchers noticed a genetic variation in some people with kuru that has been known to promote long incubation periods. They have also proposed that individuals having contracted CJD in the early s represent a distinct genetic subpopulation, with unusually short incubation periods for bovine spongiform encephalopathy BSE. This means that there may be many more vCJD patients with longer incubation periods, which may surface many years later. Large scale studies in the UK have yielded an estimated prevalence of per million, higher than the actual number of reported cases. This finding indicates a large number of asymptomatic cases and the need to monitor. It was discovered that all had consumed squirrel brains, although a coincidental relationship between the disease and this dietary practice may have been involved. From November to April , four suspected cases of the disease arose in Rochester. Researchers believe one in 2, people in the UK is a carrier of the disease linked to eating contaminated beef vCJD. This new study examined over 32, anonymous appendix samples. Of these, 16 samples were positive for abnormal prion protein, indicating an overall prevalence of per million population, or one in 2, people are likely to be carriers. No difference was seen in different birth cohorts â€"60 and â€"85 , in both sexes, and there was no apparent difference in abnormal prion prevalence in three broad geographical areas. Genetic testing of the 16 positive samples revealed a higher proportion of valine homozygous VV genotype on the codon of the gene encoding the prion protein PRNP compared with the general UK population. This also differs from the patients with vCJD, all of whom to date have been methionine homozygous MM genotype. The concern is that individuals with this VV genotype may be susceptible to developing the condition over longer incubation periods.

9: Creutzfeldt-Jakob disease | Alzheimer Society of Canada

Only a brain biopsy or an examination of brain tissue after death (autopsy) can confirm the presence of Creutzfeldt-Jakob disease. But doctors often can make an accurate diagnosis based on your medical and personal history, a neurological exam, and certain diagnostic tests.

As the disease progresses, there may be rapidly progressive deterioration of cognitive processes and memory dementia , resulting in confusion and disorientation, impairment of memory control, personality disintegration, agitation, restlessness, and other symptoms and findings. Later stages of the disease may include further loss of physical and intellectual functions, a state of unconsciousness coma , and increased susceptibility to repeated infections of the respiratory tract e. In many affected individuals, life-threatening complications may develop less than a year after the disorder becomes apparent. In approximately 90 percent of cases, CJD appears to occur randomly for no apparent reason sporadically. About 10 percent of affected individuals may have a hereditary predisposition for the disorder. Reports in the medical literature suggest that familial cases of CJD are consistent with an autosomal dominant mode of inheritance. In addition, in some extremely rare cases, CJD may take an infectious form. The disorder is thought to result from changes mutations in the gene that regulates the production of the human prion protein or direct contamination transmission with abnormal prion protein in infected brain tissue. In , experts suggested the possibility that this variant might be associated with consumption of beef from cows with a related infectious brain disorder known as Bovine Spongiform Encephalopathy BSE or "Mad Cow Disease. Later, BSE also began to appear in some other European countries. In addition, coordinated national and international efforts are in place concerning the prevention, study, and surveillance of BSE and CJD. In early December , European Union agriculture ministers agreed upon new measures to combat the spread of mad cow disease, including incinerating any cow over 30 months of age that had not tested negative for BSE. BSE is thought to become detectable and infectious when cattle are approximately 30 months old. Individuals with Creutzfeldt-Jakob disease may then experience rapidly progressive loss of intellectual abilities, demonstrating impaired memory and judgment and distinct personality changes dementia. Vision may also become increasingly impaired. In individuals with CJD, neurological and neuromuscular impairment continues to progress, and later stages of the disorder may be characterized by loss of physical and intellectual functions, coma, and increased susceptibility to repeated infections of the respiratory tract e. In many cases, life-threatening complications tend to develop less than a year after the disorder becomes apparent. V-CJD appears to occur in younger individuals i. Variant Creutzfeldt-Jakob disease appears to be initially characterized by depression, anxiety, withdrawal, and personality and behavioral changes. Delusions are sometimes reported. In some cases, individuals with the disorder may have abnormal sensations dyesthesia or pain in the face, arms, and legs. Within a few weeks or months, affected individuals experience the onset of progressive neuromuscular symptoms including an impaired ability to coordinate voluntary movement cerebellar ataxia ; severely diminished muscle tone hypotonia ; and slow, halting speech. In some cases, neuromuscular abnormalities may include irregular, rapid, involuntary jerky movements chorea. As the disease advances, individuals with V-CJD demonstrate increasing memory impairment that progresses to dementia. During later stages of the disorder, affected individuals may experience repeated, involuntary, shock-like muscle spasms myoclonus. In individuals with V-CJD, life-threatening complications tend to develop approximately two years after initial symptoms occur. Causes Scientists believe that a transmissible agent is responsible for causing Creutzfeldt-Jakob disease. Initially, this was thought to be a slow virus, since a period of many years may elapse between the initial exposure and the appearance of symptoms. However, today it is believed that this agent is very different from viruses and other known infectious agents. Instead, the agent is called a prion, and it is thought to transform normal protein molecules into infectious ones. Although the disease is caused by a transmissible agent, it is not considered to be contagious in the traditional sense. In approximately 90 percent of cases, Creutzfeldt-Jakob disease CJD appears to occur randomly for unknown reasons sporadically. About 10 percent of affected individuals may exhibit a hereditary predisposition for the disorder. In some extremely rare

cases, CJD may take an infectious form. The term prion stands for proteinaceous infectious particles. Abnormal changes in the prion protein are thought to play some role in causing deterioration in certain areas of the brain, appearing as sponge-like holes and gaps thus, the term spongiform encephalopathy. Such spongiform degeneration in turn results in the progressive neurological and neuromuscular symptoms associated with CJD. The gene that regulates the production of the human prion protein, known as prion-related protein or PRNP, has been mapped to the short arm p of chromosome 20 20ppter. Chromosomes are found in the nucleus of all body cells. They carry the genetic characteristics of each individual. Pairs of human chromosomes are numbered from 1 through 22, with an unequal 23rd pair of X and Y chromosomes for males and two X chromosomes for females. Chromosomes are further subdivided into bands that are numbered. Some researchers suggest that the normal cellular prion protein PrPc plays an essential role in preventing the degeneration and loss of brain cells. During one laboratory study, when researchers removed the normal prion protein from nerve cells i. However, when researchers restored normal PrPc to cells without the protein, affected neurons returned to health. Further research is necessary to determine the implications of such findings. Many researchers suggest that CJD and other prion diseases result from abnormal changes in the shape of the prion protein. The modified form of PrPc that may cause disease is known as PrPsc for scrapie prion protein. As a result, abnormal prion proteins gradually accumulate, forming fixed deposits plaques in the brain and the associated, progressive neurological and neuromuscular impairment seen in those with such disorders. Laboratory studies conducted by an international research team suggest that a relatively small, specific portion of the prion protein i. The researchers demonstrated that adding the specific prion peptide to cells affected by scrapie, a form of spongiform encephalopathy that originates in sheep and goats, prevented the production of new, abnormal prion protein. Such findings suggest that a specific part of the prion protein itself may be used to block its ability to fold incorrectly, potentially preventing progression to disease. However, much additional research is required before it may be determined whether such findings may have practical treatment implications in individuals with CJD or other forms of human prion disease. In March of , the British government announced that consumption of beef from cows with an infectious brain disorder Bovine Spongiform Encephalopathy may have caused a variant form of Creutzfeldt-Jakob disease V-CJD in several young people in the United Kingdom. As with CJD, abnormal changes in prion proteins are also thought to play a role in the brain degeneration associated with BSE. BSE was first recognized in the UK in and was linked to contaminated cow feed composed of sheep meat and bone meal. The sheep may have been infected with scrapie. In the late s, Britain banned the use of such feeds in animals raised for human consumption. All of the cases identified in England had a particular genetic trait, which is not a mutation, that may have predisposed them to the condition. The genetic abnormality involves a variation of the prion protein. Approximately 40 percent of the British population has this genetic trait. Creutzfeldt-Jakob disease CJD appears to affect males and females in equal numbers. It is an extremely rare human disorder that occurs worldwide with an incidence rate that has remained stable at approximately one case per million people annually. In individuals in which CJD is thought to take an infectious form e. In individuals with the classical form of Creutzfeldt-Jakob disease i. In some but not all rare cases of classical CJD in which the disorder is thought to be inherited, the disorder may become apparent earlier in life such as in the third or fourth decade. The clinical course of the disease tends to be rapidly progressive, with life-threatening complications occurring less than a year after the disorder becomes apparent. Variant CJD appears to affect primarily individuals before the age of approximately 40 years, with many cases occurring in adolescents. V-CJD appears to have a more extended clinical course, with life-threatening complications typically occurring approximately two years after initial symptoms occur. Comparison may be useful for a differential diagnosis: Prion diseases There are additional rare degenerative brain disorders that are human prion diseases. These include Gerstmann-Straussler-Scheinker disease, fatal familial insomnia, and kuru. Gerstmann-Straussler-Scheinker GSS disease is a rare degenerative brain disease that is transmitted as an autosomal dominant trait. The disorder is typically characterized by increasingly impaired coordination of voluntary movements cerebellar ataxia , with associated unsteadiness, clumsiness, imbalance, and an abnormal manner of walking gait disturbances. With disease progression, affected individuals may develop involuntary,

rhythmic, rapid eye movements nystagmus and abnormally slowed, slurred speech dysarthria. Additional findings may include stiffness rigidity , unusually slow movement bradykinesia , and, in some cases, slowly progressive deterioration of mental functioning dementia. GSS disease is caused by certain specific changes mutations in the gene i. Classic GSS disease is typically distinguished from CJD by an earlier age at symptom onset, a longer duration of disease progression, slowly evolving dementia, more prominent signs of cerebellar ataxia, and differences in degenerative changes of the brain e. However, a form of GSS disease has been described in a Hungarian family with three affected sisters in whom associated symptoms were indistinguishable from those associated with sporadic CJD. According to researchers, the implications of such findings are currently unknown. Fatal familial insomnia FFI is a rare, rapidly progressive, degenerative brain disorder that is transmitted as an autosomal dominant trait. The disorder typically becomes apparent during middle age or later life and is characterized by an inability to sleep or abnormal wakefulness that is resistant to treatment intractable insomnia and impaired functioning of the portion of the nervous system i. Dysautonomia may be characterized by fever pyrexia , profuse sweating diaphoresis , abnormal contraction of the pupils miosis , and other associated findings. These may include increased reflex responses hyperreflexia ; impaired coordination of voluntary movements ataxia ; tremors; and involuntary, shock-like contractions of certain muscles myoclonus. Neurodegenerative changes associated with FFI may be limited to certain regions of the brain e. Fatal familial insomnia is caused by a specific mutation of the PRNP gene on chromosome Kuru is a rare progressive degenerative brain disorder that occurs exclusively in members of the Fore linguistic tribal group of the New Guinea highlands. Neurodegenerative changes include generalized loss of nerve cells, particularly in the outer region of the brain cerebral cortex , and the development of characteristic plaques i. Transmission of the disease is thought to result from ritualistic handling and ingestion cannibalism of brain tissue of deceased relatives. The incidence of kuru has dramatically declined with the cessation of such practices. Neurodegenerative changes lead to the formation of plaques or patches within the brain and the loss of cholinergic neurotransmitter function. The early behavioral changes may be subtle; however, as the disease progresses, memory losses increase and there are personality, mood, and behavioral changes. There may also be disturbances of judgment, concentration, and speech along with confusion and restlessness. There are several additional progressive conditions of the brain e. For more information on these disorders, choose the exact disease name in question as your search term in the Rare Disease Database. Diagnosis According to the medical literature, Creutzfeldt-Jakob disease CJD should be considered in adults who experience a sudden onset of rapidly progressive dementia and neuromuscular symptoms such as repeated, involuntary, shock-like muscle spasms myoclonus. However, confirming a diagnosis may be difficult since other neurological disorders share similar symptoms; in addition, laboratory tests may not detect abnormalities associated with CJD. In rare cases, computer-assisted tomography CAT scanning may reveal deterioration in certain areas of the brain, findings that may be associated with a number of other neurological disorders. However, such EEG testing alone cannot provide a definitive diagnosis.

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