CONDITON: Creutzfeldt-Jakob Disease

What is it: Creutzfeldt-Jakob disease is a degenerative brain disorder that leads to dementia and death. CJD captured public attention in the 1990s when some people in the United Kingdom developed a form of the disease — variant CJD (vCJD) — after eating meat from diseased cattle. However, "classic" Creutzfeldt-Jakob disease hasn't been linked to contaminated beef. Although serious, CJD is rare, and vCJD is the least common form.

Who is at risk: 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 60. Onset of familial CJD occurs slightly earlier and vCJD has affected people at a much younger age, usually in their late 20s.
Genetics. 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.** People who've received human growth hormone derived from human pituitary glands or who've had grafts of tissue that covers the brain (dura mater) may be at risk of iatrogenic CJD.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.

Cause: 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. But when they're misshapen, they become infectious and can harm normal biological processes. The risk of CJD is low. The disease can't be transmitted through coughing or sneezing, touching or sexual contact. The three ways it develops are: **Sporadically.** Most people with classic CJD develop the disease for no apparent reason, termed spontaneous CJD/ sporadic CJD, this type accounts for the majority of cases. **By inheritance.** In the United States, about 5 to 10 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. **By contamination.** 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.

Symptoms: Rapid mental deterioration, usually within a few months. Initial signs and symptoms typically include: Personality changes, anxiety, depression, memory loss, impaired thinking, blurred vision or blindness, insomnia, difficulty speaking, difficulty swallowing, and sudden, jerky movements. As the disease progresses, mental symptoms worsen. Most people eventually lapse into a coma. Heart failure, respiratory failure, pneumonia or other infections are generally the cause of death. Death usually occurs within a year.

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 including EEG, Spinal fluid tests, and MRI.

Complications: Dementia, withdrawn from friends/family, coma, and ultimately death

Treatment: There is no known treatment, care is palliative to make patient comfortable

Did you know? Worldwide, there is an estimated one case of CJD diagnosed per million people each year, most often in older adults.

Source: MayoClinic.com