

**CHAMPION®**

Expanding Encyclopedia Of Mortuary Practices

Number 625, 1995

CREUTZFELDT-JAKOB DISEASE AND RELATED DISORDERS

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Part 1

ABSTRACT: An indepth discussion and explanation of Creutzfeldt-Jakob disease along with several related prion driven disorders are covered for the elucidation and education of the embalmer. Early history of the various diseases is outlined with the interrelationships to kuru, scrapie, BSE and other similar neuro-degenerative diseases. The bizarre nature of the causative agent is discussed in detail along with its remarkable survivability. The dangers and concerns that embalmers have are delineated and placed in perspective. A suggested protocol to minimize risk during embalming is presented. A summation and suggestions for embalmers completes the article.

INTRODUCTION: Creutzfeldt-Jakob disease is a disease of bizarre nature that almost every embalmer or funeral director has heard of but has virtually no idea concerning the facts and realities of the disease itself. There is a very small amount of information that has been available to the embalming profession concerning Creutzfeldt-Jakob disease and unfortunately it is either incomplete, misleading or outright incorrect. There is more conjecture and rumor involved with this disease than almost any other disease and there is virtually no valid information available to help the embalmer. This has resulted in a definite uncertainty and even panic in most situations involving embalmers when they encounter this disease during the course of their professional career. Nothing is more fearful than complete lack of information concerning a potentially deadly disease.

The intent of this article is to introduce you, the embalmer, to the strange world of the Human Transmissible Spongiform Encephalopathies, such as Creutzfeldt-Jakob disease and supply you with the information you need to make intelligent, informed choices when you are faced with dealing with Creutzfeldt-Jakob disease and how best to protect yourself in the situation when it arises.

EARLY HISTORY: The strange and convoluted story begins with a curious dementia case reported by Dr. Creutzfeld in 1920 in Germany. The case involved a 22 year old woman with a curious neurodegenerative disease of insidious onset and short duration followed by death. In 1921, Dr. Jakob further elaborated on this type of unusual dementing disorder by describing 4 dementia cases, all of middle-aged patients with remarkably similar symptoms and all rapidly resulting in death. These cases and others were followed through the 1930's to 1950's and were left essentially as an enigma or causation by an unknown agent or agents. Interestingly, on a bizarre note, the original case by Dr. Creutzfeldt may not have actually been the famed Creutzfeldt-Jakob disease that bears his name. There is general agreement that most if not all of the cases elucidated by Dr. Jakob were the syndrome and sophisticated analysis of some remaining tissue samples of the cases confirm Creutzfeldt-Jakob disease as the culprit.

KURU, A CANNIBAL DISEASE: The real breakthrough for understanding Creutzfeldt-Jakob disease occurred in 1957 with the discovery of a curious degenerative brain disease of cannibals. Certain Fore tribes of Papua, New Guinea were inflicted with a disease called Kuru - the trembling with fear. This disease manifested itself as a fatal disease with an onset and duration that lasted usually 1 year. To date there have been 2500+ cases reported in these relatively small villages with an occurrence of at least 1% of the population. It was prevalent in women and children but rarely affected adult males. The terminal stage of this brain disease was marked by total dementia, muteness, inability to move, ataxia (jerkiness of the muscles), dysarthria, dysphagia and death.

Kuru was determined to be linked to the ritual cannibalism that was practiced by these tribes. The brains of the elders were ritually prepared and eaten by the women and children, but seldom by the men. This endocannibalism was outlawed by the authorities of New Guinea and now the disease is slowly disappearing. There are still cases appearing due to the extreme incubation periods of the disease -exceeding 30 years in some instances. Kuru was successfully passed to chimpanzees in 1966 by intracerebral injection of brain specimens. This disease however, was not passed maternally or by mother's milk which was determined by very careful epidemiological studies. Further studies have even determined the exact cannibal feast that resulted in the transmission to certain victims. A suggested link to a bizarre disease of sheep (scrapie) was proposed in 1959 and ultimately lead to the path of understanding for Creutzfeldt-Jakob disease.

SCRAPIE - AN ANIMAL TRANSMISSIBLE SPONGIFORM ENCEPHALOPATHY: This brain degenerative disease of sheep has been known for over 200 years and usually develops in breeding ewes that are 2-4 years old. The disease takes its name from the scraping against trees and fence posts by the sheep to alleviate an intense itching with brain degeneration and death as the final result. In the 1930's a massive outbreak of scrapie was apparently caused by the use of formalin-treated louping-ill virus vaccine that destroyed entire flocks and diseased over 1500 sheep. Formaldehyde, in this situation, was proven inadequate in eliminating or neutralizing the causative agent of scrapie.

Scrapie has been transmitted to several animals by various routes such as intracerebral injection, parenteral injection and orally (with the oral route being the most unlikely, yet very possible). The agent causing scrapie has been shown to survive in soil for years and consequently, no ploughing under of sick sheep is now allowed. At first, this disease was thought to be related to visna (a retroviral true slow virus disease of sheep) but such is not the case. Visna is essentially a sheep counterpart to an AIDS-type dementia complex of humans. Despite years of research, it is still not known how scrapie spreads in sheep flocks. It is quite obvious though, that scrapie does indeed spread very insidiously and easily through sheep.

CREUTZFELDT-JAKOB DISEASE: This disease is a neurodegenerative disease that evokes no apparent immune or inflammatory response and results in dementia, cortical blindness, motor disorders, rigidity and myoclonus (muscle spasms) and some uncontrollable trembling with the result being death usually within six months of onset of symptoms. There are no documented recoveries, the disease is fatal. EEG (electroencephalographic) traces appear normal but do exhibit abnormalities later in the course of the disease. Creutzfeldt-Jakob disease occurs on a worldwide basis of 1 per million population and the peak age of occurrence is 60-65 years of age. There are, however, several examples of much earlier onset. It occurs twice as often in an urban environment as in a rural setting. There are at least 250+ diagnosed cases of Creutzfeldt-Jakob disease every year in the United States.

Approximately 15% of the cases appear to be inherited and demonstrate an autosomal dominant pattern of gene expression. The rest are sporadic with uncertainty as to the cause. There appears to be some clustering in certain ethnic groups such as Libyan Jews and Czechoslovakian herdsmen with the implication that contact with infected sheep herds and the eating of sheep brains and eyeballs may contribute to the incidence of the disease. This connection is not certain and is now suspect with some researchers who believe it is more likely due to a genetic susceptibility to the disease. Exactly how sporadic cases of Creutzfeldt-Jakob disease do occur is not known with certainty.

There has been one other unfortunate aspect of the disease - in iatrogenic cases (medically induced by treatment). There are numerous proven transmissions of the disease resulting from neurosurgical contamination of stereotaxic electrodes that were disinfected with formaldehyde, 12 cases of disease from corneal transplants, over 45 cases from pituitary extract hormone and gonadotropin from cadavers (unfortunately in most cases transmitted to children) and several cases of transmission from dura mater grafts from infected cadavers. The most bizarre case involving neurosurgery infection was the successful transmission of Creutzfeldt-Jakob disease from electrodes that were confirmed to have transmitted the disease to patients despite sterilization with formaldehyde. The electrodes were stored for three years then cleaned and sterilized with formaldehyde/ethanol three times then implanted into the brain of a chimpanzee. After eighteen months, the animal was positively diagnosed with the disease. The use of cadaver derived pituitary hormone extracts was banned in 1985.

There is no direct correlation, but approximately 30 health care workers (such as neurosurgeons, autopsy pathologists, etc.) so far have contracted the disease. This, however, does not appear to be greater than expected from the normal population but caution due to the lack of understanding of the exact causes of sporadic infection is advised. The causative agent of Creutzfeldt-Jakob disease has been found in brain, spinal cord, cerebrospinal fluid and less in lymph and lymph tissues, liver, kidney, lung, cornea and finally blood. It is not certain if it is possible to transmit by mothers milk due to two conflicting reports, but it does appear possible, but very inefficiently, at best. It was first not thought possible to transmit by blood transfusion but it now appears likely that this has occurred. Four cases of probable blood transfusion Creutzfeldt-Jakob disease have appeared in Australia where females that were treated with cadaver derived pituitary hormone for infertility subsequently donated blood. It appears quite possible from animal studies that transmission by placenta, milk and umbilical cord exists at least in lower animals. Maternal transmission in animal studies are uncertain but it is positively determined that there is fetal sharing of maternal erythrocytes.

Definite diagnosis of Creutzfeldt-Jakob disease has been uncertain until very recently where two-dimensional electrophoresis is now possible in some cases with reasonably accurate results ante-mortem. Previously, a definitive diagnosis was only possible at autopsy or at best by a dangerous brain biopsy procedure. There have been several misdiagnoses of Creutzfeldt-Jakob disease as Alzheimer's disease. Some researchers feel that due to misdiagnoses, Creutzfeldt-Jakob disease is underreported and occurs more frequently than it appears. Alzheimer's disease is also 85% sporadic and 15% familial (linked to inheritance) with an incidence of 1 out of 4 by the age of 85. The duration is typically longer with 5-10 years being the average. Alzheimer's disease is predicted to become of epidemic proportions in the coming decades as the elderly live longer and longer. Alzheimer's disease also displays numerous beta-amyloid plaques with numerous neurofibrillary tangles (these are distinct however from the plaques associated with Creutzfeldt-Jakob).

GERSTMANN-STRAUSSLER-SCHEINKER SYNDROME: This is an unusual variant of a Creutzfeldt-Jakob type disease that was first diagnosed in 1928 by Gerstmann and followed with seven additional cases in 1936 by all three researchers. It has a very similar appearance to Creutzfeldt-Jakob but differs in several important ways. It is very rare with an occurrence of 1 in 10 million of population and has an earlier onset of symptoms at approximately the 4th decade of life with a longer duration of typically 5 years. The symptoms are dementia, cerebellar ataxia and pyramidal signs (of the cerebral cortex) with death being the final result. Gerstmann-Straussler-Scheinker syndrom appears to be an inherited form of a Creutzfeldt-Jakob type disease affecting only certain families (of which several kindreds have been identified throughout the world).

CONTINUED: Creutzfeldt-Jakob Disease and Related Disorders
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