

CREUTZFELDT-JACOB DISEASE — MAD COWS AND CANNIBALS IN THE HOSPITAL CORRIDORS

by Karen C. Duncan, Registered Nurse and Attorney at Law.



The modern scientific thinking on Creutzfeldt-Jacob Disease (CJD) began with the study of the primitive Fore tribe (pronounced 4-a) in eastern New Guinea. There had long been a strange neurologic disease in the cannibalistic jungle tribe. Some of the Fore people, primarily the women and children, began to have difficulty walking. The disease then progressed into a generalized tremor, slurred speech, bizarre behavior and a fixed facial grimace resembling a smile. Called the “laughing death,” its name is Kuru. Ending in violent, uncontrolled movements, nystagmus and wasting, it was 100% fatal just as CJD is today.

The first account of this disease was published in the New England Journal of Medicine in 1957. The doctors investigating the disease found that the victims had microscopic holes in their brains, resembling those of a sponge. The investigators also realized that the disease was spread by the cannibalism, specifically the ritual eating of the brains of deceased relatives by the women and children. Only the men got to eat the prime muscle tissue.

At first, most researchers believed that the infectious agent was a virus. However, it did not act like a virus. It produced no antibody response, and it survived the heat, pressure, radiation and ultraviolet light commonly used for sterilization.

By 1982, a leading neurologist hypothesized that the infectious agent was a novel proteinaceous infectious particle or “prion”(pronounced pree-on). Although ancient, it was finally recognized as a whole new kind of disease, neither virus nor bacterium, and it was resistant to most types of sterilization because it contained no nucleic acid. Eventually, scientists linked the disease to a well known and ancient disease of sheep, “scrapie” (so-called because sheep scraped against objects to relieve the intractable itching caused by the disease).

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This “spongiform encephalopathy” (referencing the sponge-like appearance of the brain) is also found in goats, deer, elk, cattle, mink, domestic cats, and monkeys.

The human form, Creutzfeldt-Jacob Disease, is found worldwide and occurs in about one diagnosed case per million of the population. Although 95% of the cases are thought to be sporadic, the remaining 5% comprise genetic, iatrogenic or nosocomial causes. CJD normally strikes between the ages of 50 and 70 and presents as progressive mental deterioration soon associated with visual deterioration and muscle twitching. It does not induce fever and the cerebral spinal fluid looks normal in conventional tests. CT scans, MRIs, PETs, X-rays and most laboratory tests are not helpful in diagnosing CJD, so it is frequently misdiagnosed. A new test is being developed to diagnosis CJD through cerebral spinal fluid analysis but the test’s reliability is unknown.

CJD is often mistaken for a variety of other psychological and neurological diseases such as Alzheimer’s, Pick’s disease, Huntington’s disease, cerebral hematomas and vascular irregularities. A definitive diagnosis requires a brain biopsy, but even a brain biopsy may sometimes produce a false-negative if the biopsied area was unaffected by the disease. There is no treatment or prophylaxes.

The incubation period of infectious CJD can extend up to 30 years after inoculation. The recent outbreak of a variant of CJD (vCJD) in Great Brittan, caused by eating the meat of infected cattle, was identified primarily because it had a much shorter incubation period and thus developed in younger people.

CJD is not contagious in the traditional sense. A CJD patient’s family members or CJD medical caregivers have no greater risk of getting it than the general population. The primary source of infectious CJD is through infected nervous tissue (especially brain, spinal cord, and eye tissue) in direct contact with central nervous system tissue. Infectious CJD occurs mainly through implantation of contaminated electrodes in the brain, dura matter grafts, surgical instruments and the injection of human growth hormone derived from cadaveric pituitaries. The first known victim of iatrogenic CJD was the recipient of a corneal graph from a donor who had died of undiagnosed CJD.

The infectious agent of CJD, prions, exhibits unique resistance to conventional chemical and physical

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decontamination. It is not killed by most common disinfectants, tissue fixation or autoclaving conditions. It is also extremely resistant to ionizing and ultraviolet radiation. One of the world's leading experts on the disease, Dr. Paul Brown of the National Institute of Health, has said that prions are "almost immortal."

Consequently, although CJD is a rare disease, its resistance to sterilization presents an important and complex problem for hospitals. Because CJD can persist on sterilized surgical instruments and infect subsequent surgical patients, the infection control team should be immediately informed when a known CJD patient is admitted to the hospital. A written policy should be in place for the nursing areas, laboratory, pathology and the surgical areas. Specific protocols should be identified regarding preoperative, peri operative and postoperative management of the patient. For neurologic and ophthalmic surgical procedures, single use instruments should be used whenever possible. Any other instruments used on the CJD patient's neurological tissue should be destroyed. Subsequent use of stereotactic intra-cerebral needles or probes have been identified as particularly likely to transmit this infection.

CJD patients are often undiagnosed when admitted to the hospital; the admitting diagnosis could be dementia, rule out meningitis or Parkinson's. Diagnostic tests and even brain biopsies are often performed before the CJD diagnosis is made. If the surgical neurological instruments used on the CJD patients are used on subsequent patients in the interim, those subsequent patients are at risk for infection. Serious legal liability and adverse publicity could result. In this situation, hospital administration and the infection control team should immediately be notified. The subsequent, at-risk patients should be quickly identified and informed according to HCA guidelines. The surgical instruments used on the neurological tissue of the CJD patient must be isolated from use. Some hospitals that have experienced this problem recommend numbering the neurological surgical trays and recording the identifying number on the operative record. In that way, the hospital can more easily identify which tray was used on the CJD patient and thus more easily identify the subsequent uses.

The story of the Fore people has not ended. After the cannibalism ceased in the early 1960's, the incidence of the disease began to abate. However, because of its long latency period, tribal members are still showing onset of Kuru symptoms.

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Mad cow disease has now been found in several Western European countries and recently in Japan. Although it has never been detected in the U.S., some researchers have suspected that several American game hunters have contracted CJD from consumption of elk, moose and deer infected with a spongeform disorder.

Plainly, hospitals must be prepared for unique problems associated with the care of the CJD patient and the protection of the other patients. Risk managers may get further information on the proper precautions to take from the Centers for Disease Control and Prevention (<http://www.cdc.gov>), the National Institute of Health (<http://www.nih.gov/health>). The fascinating story of the Fore people is found in *Deadly Feasts: Tracking the Secrets of a Terrifying New Plague* by Richard Rhodes.