Anthropological Perspectives on
Transmissible Spongiform Encephalopathies

Children with Kuru, advanced stage. The child on the left can no longer sit up without support. 1957.

Epidemiology

KURU is one of the Prion diseases, a slow growing, fatal neurological degenerative disease. It is highly contagious. Once infected, the individual may be asymptomatic for anywhere from a few months to thirty years. However, once symptoms begin to manifest, death follows within six months.

The earliest cases of Kuru were found among the Fore tribe -- an agricultural people numbering about 20,000 in the Eastern Highlands Province of Papua New Guinea. In the mid-1950s Kuru reached epidemic proportions, resulting in about 3,000 deaths over a twenty year period. Subsequent outbreaks of Kuru arose in artificial environments: 60 people associated with a research lab died after being infected by contaminated surgical instruments, while 85 people died after receiving growth hormone injections contaminated by prions.

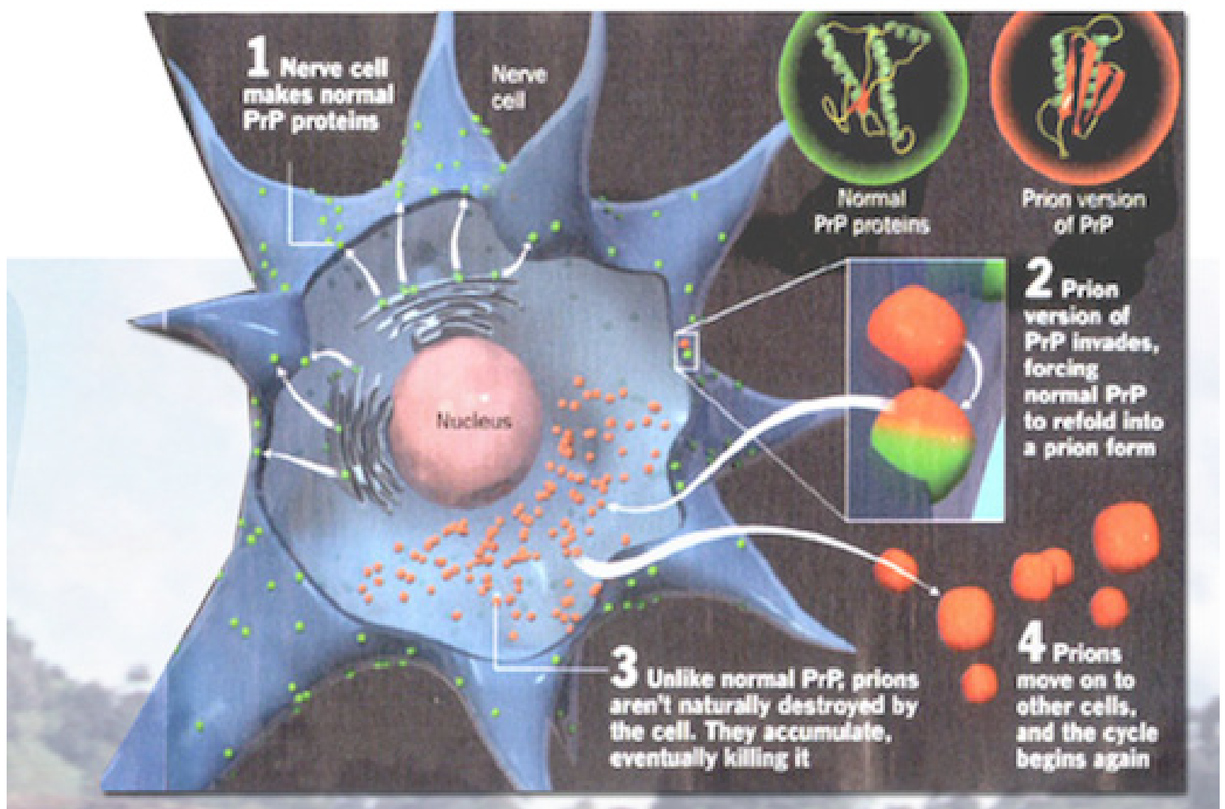
Center for Disease Control. Prion Diseases. 2010 http://www.cdc.gov/ncidod/dvrd/prions/.

Collinge J, Apers MP. Reminiscences and reflections on kuru, personal and scientific. Philosophical transactions of the Royal Society 2008(363):3613 ff.

The 1950s saw the peak of the Kuru epidemic; some villages in the Eastern Highlands of Papua New Guinea had infection rates of 20 percent and over. Because there is no treatment for this progressive neurological disease, death is inevitable.

While Kuru seems to have died out among the Fore, closely related Prion diseases (called Transmissible Spongiform Encephalopathies) still cost lives. The most prevalent of these is bovine spongiform encephalopathy (mad cow disease) in cattle, which is transmitted to humans by ingestion of food contaminated with infected cattle tissues.

A epidemic of BSE in England in the 1990s caused approximately 300 human deaths and the destruction of tens of thousands of cattle.



Symptoms and Stages

ambulant stage: unsteadiness gait, voice, hands, and eyes; deterioration of speech; tremor; shivering

sedentary stage: no longer walk without support, involuntary movements (choreoathetosis, myoclonic jerks, fasciculations), emotional lability, outbursts of laughter, depression, hallucination

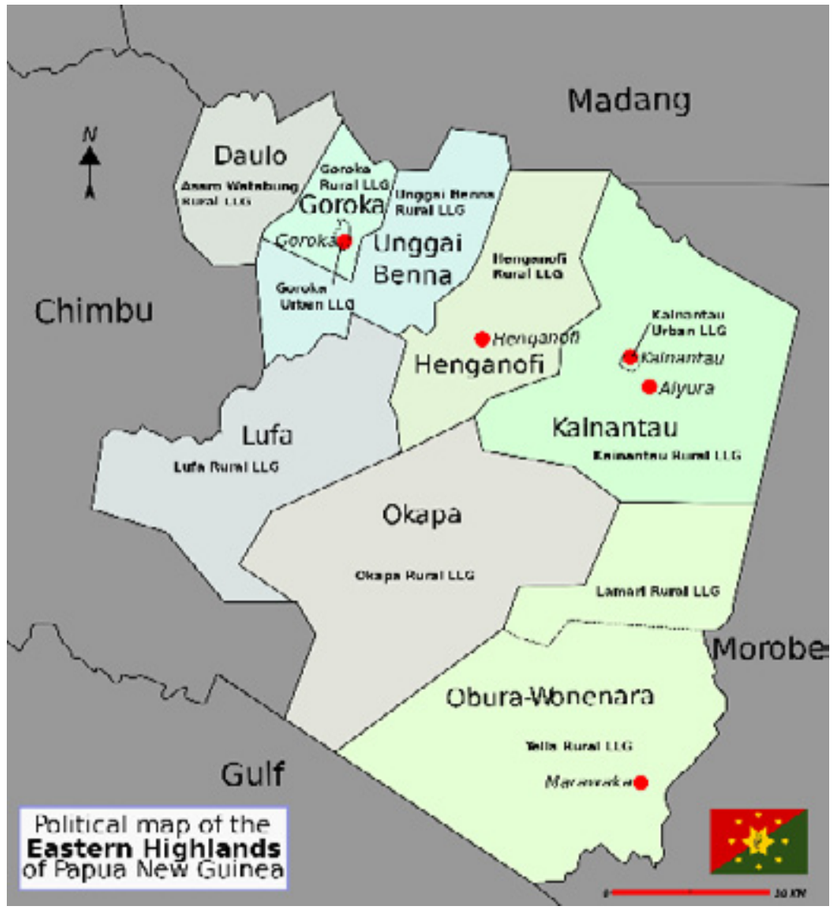
terminal: inability to sit up without support; worsening cerebellar ataxia, tremor and slurring of speech; urinary and faecal incontinence; difficulty swallowing (dysphagia); deep ulcerations, dementia

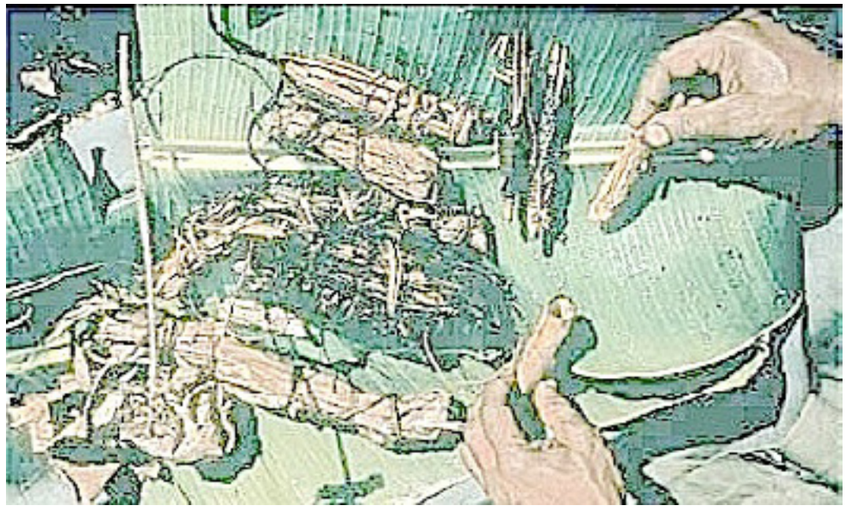
Viral Disease

A proteinaceous infectious particle, or prion (PREE-on) consists of protein alone. Prions are the only known example of infectious pathogens that are devoid of nucleic acid. They propagate by transmitting a mis-folded protein state that the protein cannot replicate by itself. This induces pre-existing normal forms of the protein to convert into the rogue form (the process by which prions stimulate the conversion of PrP(C) to PrP(Sc) is not clear). New prions can then go on to convert more proteins themselves triggering a chain reaction. Abnormal prion proteins PrP(Sc) accumulate in neurons normal immune protective processes ineffective.

prions completely clog the infected brain cells, which misfire, work poorly, or don't work at all.
Infected prion-bloated brain cells die and the released prions then enter, infect, and destroy other brain cells. At death the brain is riddled with holes and looks sponge-like..

References and further reading can be found on the class website.





Above: At the height of the epidemic, Moke and Miarasa villages met to disclose and destroy sorcerers' magic disease-producing packages (see here) for the three diseases of kuru, tukabu and analisa, all believed to be the result of sorcery.

Among the Fore, Kuru was transmitted by ritual mortuary cannibalism.

Upon the death of an individual, the maternal kin would remove the arms and feet, strip the limbs of muscle, remove the brains, and cut open the chest in order to remove internal organs. Lindenbaum (1979) states that kuru victims were highly regarded as sources of food, because the layer of fat on victims who died quickly resembled pork. Women also were known to feed bits of brain and various parts of organs to their children and the elderly. Only 2 percent of Kuru victims in PNG are male.

Well into the 1960s government attempts to stop ritual cannibalism met with resistance. Within the tribes it was believed that sorcery was the cause of Kuru, and that to abandon the religious ritual of cannibalism would make the situation worse.

Individuals hid their dead until it was safe to retrieve a body and proceed with the ritual preparation, without fear of interference from authorities.

"The disease was researched by Daniel Carleton Gajdusek as part of an international collaboration with Australian doctor [...]] In the mid-1960s Alpers collected post-mortem brain tissue samples from an 11-year-old Fore girl, Kigea, who had died of kuru. He took these samples to Gajdusek in the USA, who then injected two chimpanzees with the infected material. Within two years, one of the chimps, Daisy, had developed kuru, demonstrating that the unknown disease factor was transmitted through infected biomaterial and that it was capable of crossing the species barrier to other primates."

Collinge J, Whitfield J, McKintosh E, et al. (June 2006). "Kuru in the 21st century—an acquired human prion disease with very long incubation periods". Lancet 367 (9528): 2068–74.

Healthier days for the Fore tribe.