Kuru: a medical mystery

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Part 1: The discovery of kuru

In January 1954, an Australian administrator exploring the provinces of Papua New Guinea filed a report detailing a new disease

> The first sign of impending death is a general debility which is followed by general weakness and inability to stand. The victim retires to her house. She is able to take a little nourishment but suffers from violent shivering. The next stage is that the victim lies down in the house and cannot take nourishment, and death eventually ensues.

By 1959, it was realized that this disease was wide-spread among the Fore tribe. Study and treatment of the disease was made difficult as the Fore lived in remote conditions. The same report also noted that warfare among villages was common, the villagers believed in sorcery and performed ritual cannibalism.

Around this time an anthropologist, Shirley Lindenbaum, began to work with an Australian medical student, Michael Alpers, and an American research, Carleton Gajdusek, to study the disease.

Imagine that you are part of this team, entering Papua New Guinea to study a new disease. What theories do you have about the potential cause of the disease?

What evidence would you need to test your theory?

Part 2: experimental evidence

In 1959, a veterinary neuropathologist William J. Hadlow happened to hear about kuru from a colleague at dinner (see - socializing is important for science!). He immediately saw similiarties between *kuru* and the disease scrapie he was studying in sheep. He published a letter on this in the prestigious journal the *Lancet*, which motivated Gajdusek and Alpers to investigate the transmissability of kuru. Alpers collected brain tissue, with consent, from a recently deceased victim, and transported it to the lab where it was injected into a chimpanzee. After two years, the first symptoms of *kuru* begain to show in the chimps.

What are the implications in showing that *kuru* is transmittable from one victim to another? Which of your theories does this support or reject? Specifically, is this evidence compatible with a genetic basis for kuru among the Fore people?

Part 3: epidemiological evidence

While waiting for the results of the transmission experiment, Alpers reviewed the epidemiological data for *kuru*. Two patterns emerged. First, *kuru* was almost exclusively restricted to the Fore, though they came into regular contact with neighboring tribes (see map).

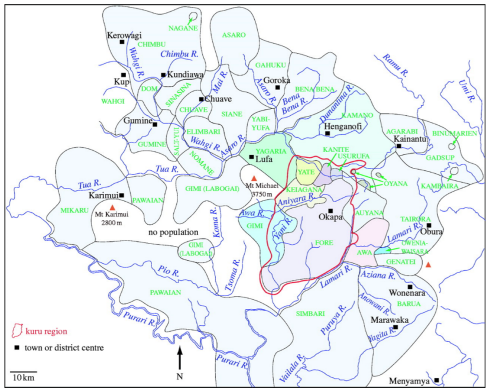

Figure 1: Map showing linguistic groups of Papua New Guinea, with kuru infected region circled in red. Alpers et al 2008 Phil. Trans. Roy Soc 363: 3707-3713

What are the implications in showing that *kuru* was restricted to one tribe? Specifically, is this evidence compatible with a bacterial or viral epidemic?

Second, Alpers graphed the data on *kuru* deaths (Figure 2). He noted that since 1960, the number of *kuru* deaths was dramatically declining, especially among the women and children. Intriguingly, this was around the same time that Australian administrators banned the practice of ritual cannibalism. The ritual cannibalism had been performed primarily by the women and children.

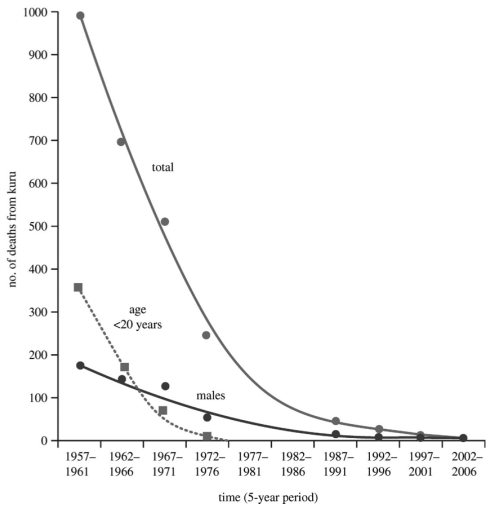

Figure 2: Death rate of the kuru epidemic by year, divided by age and gender. Alpers et al 2008 Phil. Trans. Roy Soc 363: 3707-3713

The experimental and epidemiological evidence all point to the consumption of brain tissue as the cause of *kuru*, yet there was no evidence for a bacterial or viral infection. What other biological process could be responsible for this neurological disease?

References

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