Carleton Gajdusek & Kuru

by Pierre Paul Gros

Visuals || Tex

A Remote People with a Strange Disease

Imagine perusing the November 11 issue of *Time* magazine in 1957 and reading the following in the Medicine section under the headline "The Laughing Death":

> In the eastern highlands of New Guinea, sudden bursts of maniacal laughter shrilled through the walls of many a circular, windowless grass hut, echoing through the surrounding jungle. Sometimes, instead of the roaring laughter, there might be a fit of giggling. When a tribesman looked into such a hut, he saw no cause for merriment. The laugher was lying ill, exhausted by his guffaws, his face now an expressionless mask. He had no idea that he had laughed, let alone why. New Guinea's Fore (pronounced foray) tribe was afflicted by a deadly foe. It was kuru, the laughing death, a creeping horror hitherto unknown to medicine.

[Optional video: 1963 film of young kuru patient. (2:12)]
[Optional video: Introduction to discovery of kuru patients. (1:07)]

This was the dramatic circumstance encountered by Westerners who had been monitoring the uncivilized regions of New Guinea. Kuru, the name given to the condition by the natives, meant "shaking" or "trembling." For the local Fore people, it was due to vengeful sorcery. For young American physician D. Carleton Gajdusek (pronounced Guy-do-check), it was an unknown disease that he felt ready to investigate. Ultimately, he would spend the next several decades studying this disease.

Papua New Guinea is located in the South Pacific, just north of Australia, which then administered the territory. The interior of the island had been uncharted and uncolonized until the years following World War II. Exploration of this wild country was led by governmetn patrol officers, missionaries, gold prospectors, anthropologists, agricultural officers and medical doctors from Australia. They encountered many new peoples and cultures. Among these was the Fore people, found in the southern portion of the Eastern Highlands.

The Fore presented a novel opportunity to study an "ancient" or "uncivilized" population. The Fore lived in quasi pre-historic conditions: thatched huts, rudimentary dress, no metal tools or implements. There were no hospitals of any kind. Medicine was practiced by local healers using herbal remedies. The Fore were surrounded by other communities that constantly fought and warred with each other.

[Optional video C: Fore culture, including basket-weaving and children's toys. 1957-1963 archival footage by Gajdusek. (2:18)]

Explorers of the region were especially fascinated by one aspect of Fore culture: a ritual form of mortuary cannibalism. The Fore believed that by eating their dead, they incorporated their loved ones into themselves and thus could lessen the loss and sorrow. In Fore culture, virtually every part of the deceased was consumed. Immediate family members consumed organs that were considered "powerful," such as the genitals or brains. Other organs or body parts, such as ground up bones or marrow, were subsequently distributed to more distant family and others. To the incoming explorers, this was a frightening prospect, as reported by colonial patrol officer R. R Hallivand in 1953:

> . . . the European members, and the well-fed police escort, were greeted by native men who rubbed their hands over arms and legs making enthusiastic noises as they did so . . . accompanied a motion of conveying the flesh of the leg and arm to the mouth and chewing eagerly . . . the highest degree of commendation bestowed by the natives this remark: "I like you so much I could eat you." (quoted in Anderson, 2008, p.15)

The fears proved misplaced. The Fore were quite friendly.

Anthropologists were also fascinated by the practice of "sorcery" and by the fear and respect it engendered among the Fore. The Fore believed that illness and death stemmed primarily through conscious agents: sorcery wielded from rival clans and enemies. This seemed to explain the constant warring. One form of sorcery prominent within the community was kuru, a curse which afflicted its victim with shaking, uncontrollable tremors, and death within a year. To produce kuru, sorcerers would take material closely associated with the victim, such as hair, skin, discarded food or feces, and roll it in some leaves with a stone. The rate of decay of the resultant bundle would then determine the rate of the victim's condition, with death occurring when the bundle was completely decomposed.

[Optional video D: Fore sorcery. (2:23)]

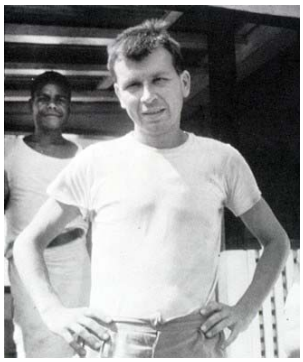
For the Fore, Westerners were just as striking, especially with their pale skin. Especially in the early years of context, many considered Westerners to be ghosts of former Fore people. In one village, a woman asked a female visitor if she had been born in the usual manner or had arrived on Earth just as she appeared (Reid 2008).

For the next several decades, the Papua New Guinea colony gained infrastructure. Departments of state were created. By the late 1950s, further exploration and colonization occurred regularly. Patrols, consisting of military officers, anthropologists and medical experts, routinely surveyed the Fore's territory. They started trading with the people: such things as salt, knives and matches in exchange for food. Colonial administrators gradually became accustomed to the Fore's sorcery.

[Optional video E: On patrol in the PNG Highlands. Archival footage, 1957-1963 by Gajudsek. (3:17)]

However, one of the patrol officers was so alarmed and puzzled by kuru that he alerted the district medical officer, Vin Zigas. Initially, Zigas considered the condition a form of hysteria, produced by the alleged sorcery. Yet he investigated, and after observations of the Fore, became convinced in 1956 that it was a disease, possibly a form of encephalitis. That meant further work in characterizing the disease and its transmission before one could hope to stem what seemed to be a growing epidemic.

Mounting an Investigation



In March, 1957, Vigas met Carleton Gajdusek. Gajdusek had been traveling the globe investigating tropical diseases, as well as studying child development in different regions and ethnic groups. Unknown diseases posed a threat to American troops stationed abroad and the U.S. National Institutes of Health (NIH) and Gajdusek collected blood serum samples from isolated populations to help them study such diseases. For Gajdusek, this allowed him to pursue research while living in the wilderness and interacting with foreign cultures. "I was interested in people that hadn't been brainwashed. . . . I needed to know cultures that had nothing to do nothing with monotheism," he later remarked (Lindquist 2009). Over the past three years he had been to Iran, Afghanistan, Turkey and, most recently, Australia, where he had collected samples from the Aborigines. Gajdusek stopped in Papua New Guinea. During conversations with the Director of Public Health there, he learned of the kuru problem and Zigas's emerging investigations. With the support of the NIH, Gajdusek turned his primary attention to kuru.

> **THINK (1)**. Imagine you are working with Zigas and Gajdusek. You do not yet know what causes kuru, so it is not yet clear how to proceed. What are the possible causes of any disease, and what type of information is relevant to confirming each? What other clues might you collect that will help indicate what type of disease kuru is?
>
> **THINK (2)**. Your investigation involves a disease among people in a vastly different culture, who do not understand Western medicine or research. What challenges might you possibly encounter, and how might you prepare for them?


Lucy Hamilton Reid had been studying nutrition in Papua New Guinea for six years. She wanted to check the diet and began a thorough survey of what the Fore were eating and how the food was prepared, including the leaves in which the food was cooked and served (Reid 2008). Patrol Officer Jack Baker thought the disease was diet, too: but something transmitted by the cannibalism! That suggestion did not fit with what was known about physiology, however, and the glib assumption -- shared by many locals -- only irritated Gajdusek, with his professional training.

[Optional video F: Fore food and cooking practices, 1957-1963. Archival footage by Gajdusek.(1:32)]

With kuru now his exclusive focus, Gajdusek took the lead in the investigation. He and Zigas aimed to characterize the disease in terms of its symptoms, distinctive features, primary treatments, and epidemiology (or how the disease was distributed by age, sex, geographical area, season, etc.). In order to complete his preliminary study, Gajdusek needed to: (a) conduct autopsies on victims of kuru, (b) collect biological samples, such as blood, feces and serum from both healthy and sick individuals, and finally (c) journey through the countryside of the Eastern Highlands to secure an accurate count of cases in the region.

Gajdusek's personal life experiences proved invaluable for the tasks at hand. During his youth, Carleton went on several nature walks and aided his aunt, who was an entomologist, as she collected insect specimens. In time, he became an avid outdoorsman and readily spent months in secluded wilderness areas. His predisposition to sleeping and trekking in the wild afforded Gajdusek the ability to complete the long walks between Fore villages and to sleep in tents or rudimentary patrol huts. His fondness for the wilderness had also served him well in his earlier work.

Gajdusek had become a master at interacting with isolated communities in Iran, Afghanistan and Australia. Growing up, Gadusek had felt an outsider. "I never felt I wanted to be like anyone I knew," he recalled later (Lindquist 2009). Now he felt that he could fit in with other cultures. At first, social barriers and fear of foreigners would limit his interaction with local tribes. But he persevered and was able to adapt to different social norms. He also developed strategies for forging fruitful relationships with native people. Most important, perhaps, he learned to interact initially with the children, who were typically less wary of outsiders. Gaining their trust, while demonstrating to others that he was not a threat, he was then able to approach adults.

Collection of biological samples was centrally important to the study of kuru. In medicine, especially in investigative epidemiology, biological samples from infected and healthy patients allow researchers to look for clues about what is causing a disease and how it is making people sick. For example, blood serum contains antibodies that can be identified in a laboratory, indicating which pathogens a person is currently battling or has encountered previously. The number and types of white blood cells can indicate if a patient is in the midst of responding to an invading pathogen or

parasite. In a case such as kuru, where the disease is uncharacterized, examination of whole organs from deceased victims may exhibit characteristic scarring or lesions, or indicate which organ system the disease affects. Finally, all these samples, in addition to feces and urine, can be used to try and isolate the causal agent of the disease — microorganisms, parasites or toxins — or their degraded byproducts.

Efforts by Gajdusek and Zigas to collect specimens from the Fore largely succeeded initially due to claims that they would cure the disease. They indicated to the natives that they expected to distribute antibiotics and other therapeutic agents to patients. However, with the lack of any relief to kuru sufferers, over the years the Fore become increasingly reluctant to trust the doctors. They also believed that the samples might be subject to further sorcery. One young Fore assistant later recalled:

> Collecting human samples was very hard owing to the fear of sorcery: the people feared that we might misplace some of the samples and sorcerers might pick them up. (Tasa 2008)

Gajdusek also wanted to conduct autopsies of kuru victims in a nearby hopspital. But the Fore custom was to bury the dead in their home ground (by this time, local officials had banned cannibalism). This proved quite a stumbling block. For all these reasons, the collection of samples and conducting of autopsies was hindered.

> **THINK (3)**. Ideally, perhaps, the Fore people should make an informed choice about participating in the medical research. Yet can they know enough about Western medicine to understand the critical importance of the samples and diagnostic practices? If not, what is an ethically warranted alternative? As Gajdusek, you have the opportunity to offer valued items to the Fore in return for blood samples and permissions to perform autopsies. What reasons would you consider for and against this option? How does this alternative respect or fail to respect the Fore people and their autonomy to make their own decisions? In what ways, if any, is the goal of scientific knowledge relevant?

Gajdusek had learned what the Fore value: matches, salt and metal tools, such as knives and axes. Drawing on his earlier experiences, Gajdusek started to "trade" these with the Fore for serum, blood, and permission to conduct autopsies. Gajdusek also observed the neurological symptoms of the disease (tremors, lack of bowel control, slurred speech) and became interested in obtained samples of full brains of individuals killed by kuru. These, too, he obtained by trading.

In order to increase his standing in Fore society, and gain greater and easier access to cadavers and brains, Gajdusek drew on his understanding of local customs. He befriended local boys, whom he then recruited into his research effort. In this region of New Guinea, boys would often wander to other villages, where they would live for a while with a

foster family and, some time later, return to their original family. In some cases, of course, children had been orphaned by parents who died of kuru. Gajdusek saw an opportunity to "adopt" Fore children as a way to integrate himself more fully into the local culture and develop further trust among the natives.

> **THINK (4)**. As a Western colleague in Papua New Guinea, what perspectives would you give to Gajdusek to consider if he proposed this possibility?

Gajdusek had never fit in comfortably in American society. An outsider all his life, when Gajdusek encountered the Fore people, he felt at ease. He wrote home to his mother indicating that he found a great affinity and rapport with the "Black primitives" and that he was enthralled (Lindquist 2009). His rapport with the young boys helped him integrate into the society and feel an intimate connection to the Fore people. Several years later (in 1963) Gajdusek began "adopting" Fore children, most of them boys.

Gajdusek's experience was not without personal difficulties. The most striking example involved a boy named Kageinaro. As Gajdusek continued his research into the 1960s, he traveled many times between America and the Fore region. Upon returning to New Guinea one time, he found Kageinaro struggling with speech and showing the primary symptoms of kuru. As death approached, Gajdusek reflected about the possibility of conducting an autopsy on the teenager:

> …how many boys will pass to a similar fate? …Starving and thirsting to death in a hideous state. It is a curse of such magnitude … that I am ashamed of my feeble efforts and appalled by my own callousness. (Anderson 2008, p. 114)

As Gajdusek collected specimens, his main focus was to characterize kuru. Samples were analyzed in three settings. Gajdusek conducted some of his research in the field in rudimentary facilities, or "bush laboratories," set up in village huts or ranger stations. These bush labs had crudely erected operating tables and a few shelves. Autopsies of kuru patients were routinely conducted in this type of setting. Gajdusek sent most samples back to the laboratories at the NIH outside Washington, D.C. for analysis. Others went to the capital city, Port Moresby, or Australia.

From the Field to News Headlines

After about 3 months of intensive work — walking between villages, mapping the disease, conducting exams, interviews and autopsies, and collecting samples and analyzing them — in late 1957 Gajdusek and Zigas published their first description of kuru. Kuru appeared to be a neurodegenerative disease exhibiting loss of motor control, such as uncontrollable tremor, incontinence, and slurred and lost speech. The disease was fatal. Treatment with steroids or antibiotics had proven ineffective: namely, it did not seem bacterial. Nor did the disease exhibit

signs of inflammation: that is, with no immune response, it seemed not to be either bacterial or viral. The brains of the victims became spongy. The illness was mainly restricted to a distinct region of Papua New Guinea. The disease afflicted and killed 1% of the population annually (about 100 victims in a population of 10,000). The disease did not strike everyone equally. Women and children were more likely to succumb. The pair had found instances within families, but the Fore were so interrelated, that it was nearly impossible to establish if it was truly genetic.

> **THINK (5)**. Develop at least one plausible explanation for why kuru was more frequent among women and children. What further observations would you seek to confirm or disconfirm this hypothesis?

The publication of the paper had several impacts. First, one prominent Australian scientist felt that Gajdusek was working in "their" territory and was now spurred further to try to remove him from the field. It was also now clear that the natives' condition, which they had earlier dismissed as psychological, was instead a significant new disease. The prospect of discovering its cause offered the potential for scientific renown. Henceforth, one could expect competition among researchers for human subjects and the precious samples. Gajdusek and Zigas tried to carry on with their research undisturbed.

Another result from the publication of the field data was stories in the mainstream press. The nature of the disease was mysterious and, coming from a remote region, exotic. They capitalized on Gajdusek's and Zigas's desciption of the slow degeneration of the patient's nervous system:

> Patients often display marked emotionalism, with excessive hilarity, uproarious, foolish laughter on slight provocation … (Gajdusek and Zigas, 1957, pp. 974-975)

The media referred to kuru as the "laughing death" and sensationalized it in their accounts, as illustrated in the opening quote.

> **THINK (6)**. Read the full 1957 article from *Time* magazine. List the significant differences between the case as presented in the article and as described in the history here. Note also aspects of the episode that seem very well presented. Discuss the significance of the similarities and differences for the typical reader of *Time*. Identify phrases that illustrate assumptions about science and the Fore culture.
>
> **OPTIONAL ACTIVITY**
> Draft a letter to the Editor of *Time* based on your analysis of the article, taking the role of either Gajdusek, Zigas, or an ordinary American reader in 1957.

"Yes, But...": Cause and Transmission

With the symptoms and distribution of the disease characterized, two major problems remained: what actually caused the disease and how was it transmitted? Evidence did not indicate clearly that it was either viral, genetic, or bacterial in nature. Indeed, Gajudsek was quite unsure if it was any of these. Meanwhile, Lucy Reid's work had helped rule out diet as a cause. That left reconsideration of Zigas's earlier idea that the disease was psychological, due to the Fore's intense belief in and fear of sorcery. The lack of apparent pathology other than neurological degeneration was puzzling. Moreover, if the disease was infectious, or transmissible between individuals, why had no Westerner yet succumbed to the disease?

Gajdusek nonetheless decided to investigate the transmissibility of the disease. For this, he needed other resources: lab animals that could be inoculated with potentially deadly material. Gajdusek collaborated with the NIH back in the United States. The experiments were fairly simple: take samples from kuru patients, suspend them in a simple liquid medium, and inject them into the animal. Then wait for signs of the disease to appear. However, several decisions had to be made first, as every trial placed an animal at risk. First, should the experiment use mice, monkeys, or both? Mice are less expensive to maintain, but monkeys are more likely to have a physiology similar to humans. Are there other values associated with each animal to consider? Second, from which organ(s) should one take the samples of kuru patients? Blood is standard, as it circulates throughout the body. Yet the disease is neurodegenerative, so the agent may be in the brain. It might also be found in the liver, responsible for so much body biochemistry.

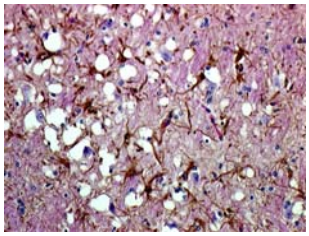
> **THINK (7)**. Develop a strategy for determining which animal(s) to use, with what kinds of samples from the kuru patients, and how many of each. Describe and discuss the scientific and ethical criteria that you use, and how you integrate them.

Gajdusek used suspensions from brains, livers and blood of kuru victims. The experiments lasted quite a long time, as the incubation period of the disease was unknown. In this case, that meant a few months (rather than a few weeks). Most test subjects were then sacrificed without much observed progress in disease development. There was no evidence of transmission, at least within that time period. Tuberculosis was known to have an incubation period of up to a few years — but this was a notable exception.

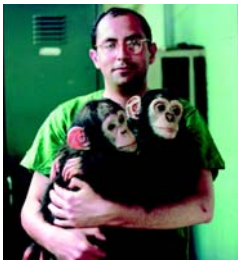
> **THINK (8)**. With the failure of these experiments to provide definitive results, what would you do next? Abandon transmission studies? Try again? (If so, what would you try differently?) What alternative explanations for kuru would you pursue, and how?

As Gajdusek started the transmission experiments, other scientists began looking more closely into the genetics of kuru. Yet the frequency of kuru

was notably higher among females. One group of researchers had proposed that a gene was expressed in females with a single copy (allele), but in males only when two copies were present. This seemed to fit the distribution of cases, but not any known pattern of how genes worked.



The initial findings on kuru also reached the attention of William Hadlow, an American veterinary pathologist working in England. He was mainly interested in a sheep disease called scrapie. A visiting friend had alerted him to a museum exhibit on kuru, another neurological disease. What caught Hadlow's notice was the similar symptoms of both scrapie and kuru: motor control debilitation and a spongy brain — and highly distinctive microscopic malformations in the brain's nerve cells (Hadlow 2008). They had been able to transfer scrapie between sheep and eventually from sheep to goats. But it took months to years. The disease agent itself had yet to be discovered. Hadlow could only characterize it as a mysterious "slow virus" (a term from an Icelandic veterinarian). Hadlow wrote to Gajdusek in 1959, telling him about the apparent similarities between scrapie and kuru. He also informed him that transfers of scrapie were often quite long and were best achieved by injecting a suspension of the brain from an infected individual directly into the brain of a healthy subject.



With Hadlow's information, Gajdusek realized that their first transmission experiments might have been too short, and that he certainly could not effectively rule out transmission. In 1963, he and Joe Gibbs at NIH began a new series of infection experiments, this time guided by the prospectively relevant findings in veterinary science. They collaborated with Michael Alpers, a physician from Papua New Guinea, who secured samples from kuru victims and then studied the animals clinically. They injected suspensions from the brains of 7 deceased kuru patients into several chimpanzees. This time, they planned to follow the animals for 10 years. Gajdusek was on a field visit to Papua New Guinea over 1½ years later when he received the news by telegram.

One of the chimps had developed symptoms of kuru-like illness. He was on the next flight back to the U.S. (Alpers 2008). Ultimately, three chimpanzees (named Georgette, Daisey and Joanna) seemed to manifest the disease, with symptoms never before observed in these primates. The symptoms appeared 18, 20 and 21 months after infection: quite an incubation period! To verify these astonishing results, they re-infected another set of chimps. In these infections, however, the initial material came, not from an infected human brain, but from the deceased chimps in the previous experiments. Results once again showed that kuru was transmissible. Most striking, for Gajdusek, was that in all the experiments, each chimp had been infected from a separate source. This, he claimed, unequivocally demonstrated the transmissibility of the syndrome. But whatever the agent was, it was unprecedented. There was no inflammation and no immunological response (evident from the autopsies of patients and primates). In addition, the delay between infection on onset of the disease was exceptionally long. Gajdusek adopted the label "slow virus."

The discovery "concerning new mechanisms for the origin and dissemination of infectious diseases" would later earn Gajdusek a share in the 1976 Nobel Prize in Medicine (Nobel Foundation, 1976).

At this point, Gajdusek had apparently solved the problem of a causal agent, although it had not yet been isolated. But the problem of transmission remained. The unidentified slow virus had to enter into the body of another individual somehow.

> **THINK (9)**. What alternatives are possible? What information would you now collect, or what experiments would you propose to investigate each?

The clear demonstration of the transmissibility of kuru was of particular interest to a married team of anthropologists, Bob and Shirley Glasse, who had been studying the Fore since 1961. Their ethnographic data was now especially relevant. By interacting with the Fore for an extended period, they had been able to track the historical spread of kuru from one initial location. That had likewise concluded that kuru seemed transmissible, not genetic. They had also suspected that it might be transmitted through cannibalistic practices. In particular, they had learned through their interviews that the ritual primarily involved women, with the assistance of children. This could account for the unequal gender and age distribution of the disease. Because the ritual involved primarily kin, kuru exhibited patterns that would misleadingly appear genetic. They could now place all their information in the context of the new findings about the nature of the transmission (such as brain tissue) and the time horizon. The ethnographic approach was continued by John Mathews, first inspired as a medical student, who tracked many individual instances of kuru to particular cannibalistic events, sometimes several years earlier. Kuru, they all concluded, was transmitted through the consumption of deceased humans (Mathews et al 1968; Lindenbaum 2008). Jack Baker's earlier hunch, it turned out, while based only on his experience as a Patrol Officer, had ultimately proven correct.

Gajdusek, however, was skeptical. He concurred that cannibalism was probably responsible, but for a different reason:

> In the cannibalism ritual, holding brain tissue, getting it on hands, and scratching mosquito bites or scabies is a much more likely method of inoculation. Since they rarely washed their hands and often picked their noses and rubbed their eyes, this is undoubtedly how they got it. (Moseley 1986).

In further experiments, colleagues at the NIH demonstrated that, certainly in the case of another disease, a slow-virus could indeed be transmitted through consuming contaminated organs. But it occurred only in spider monkeys and hamsters, never in chimps. In the absence of stronger

evidence, Gajdusek remained cautious.

A definitive answer on kuru may never be known. The Fore practice of cannibalism had officially been banned when Gajdusek and others began their investigations in the late 1950s. While some continued surreptitiously for over a decade, cannibalism had ceased by the early 1960s -- ironically, for reasons other then health considerations. Still, cases of kuru continued to appear as the latent agent became active. The last known case of kuru (of more than 2,700 total) emerged in 2002, in one of the Fore who had assisted the early research teams: incubation had taken over 4 decades. Ironically, all the research on kuru, while important to characterizing a new form of disease, ultimately contributed nothing to actually curing it or treating it among the Fore people. Fore beliefs have not changed. Most still regard sorcery as the genesis of kuru (Mathews 2008).

Epilog: From "Slow Virus" to Prion

The challenge of isolating and identifying Gajdusek's "slow virus" remained and its story was another episode of bafflement and surprise. It began in 1972 when a medical intern, Stanley Prusiner, encountered a patient with another rare, slow-virus illness, Creutzfeldt–Jakob disease. It fascinated him:

> The amazing properties of the presumed causative "slow virus" captivated my imagination and I began to think that defining the molecular structure of this elusive agent might be a wonderful research project. (Prusiner 1997)

Two years later he began work on scrapie in sheep — a pragmatic choice, since it did not involve human subjects. Still, Prusiner was interested in learning more about kuru. In 1978 he visited Papua New Guinea, trekked over the steep terrain to see several patients, and met both Gajdusek and Zigas (Prusiner 2008). Shortly thereafter, Prusiner's lab succeeded in its isolation project:

> I had anticipated that the purified scrapie agent would turn out to be a small virus and was puzzled when the data kept telling me that our preparations contained protein but not nucleic acid. (Prusiner 1997)

All known viruses then were either DNA or RNA, yet here they were apparently made of protein! That seemed impossible. Normal proteins, Prusiner eventually concluded, adopted a second shape that interfered with cell function and, more dramatically, could alter the shape of other normal proteins. Prusiner called the unusual rogue proteins "prions" (pronounced PREE-on). Gadjusek, then collaborating on a paper with him, apparently did not approve of the term (Prusiner 2008). The notion of a protein as an infectious agent was vigorously rejected by others for many years. Prusiner recalled:

> While it is quite reasonable for scientists to be skeptical of new

> ideas that do not fit within the accepted realm of scientific knowledge, the best science often emerges from situations where results carefully obtained do not fit within the accepted paradigms. (Prusiner 1997)

In 1997 Prusiner received a Nobel Prize for characterizing Gajdusek's and others' "slow viruses" as prions. While prion diseases are generally rare, they can nonetheless be quite significant.

A prion disease in cattle — bovine spongiform encephalopathy, or BSE, also known as "mad cow disease" — has on occasions threatened food supplies. As recently as 2008 the U.S. Department of Agriculture recalled 143 million pounds of frozen beef. It is not only cannibals, it seems, who may benefit from knowing about prion diseases.

THINK (10). Review the case and note examples where scientists drew on information from different fields. Describe how research was affected by such interdisciplinary links.

NOS Reflection Questions
[These are not merely review, but function to help complete and consolidate the lessons of the case study.]

Discuss how the case of Carelton Gajdusek & Kuru illustrates the following features of the nature of science:

- posing problems
- research ethics
- science journalism
- interdisciplinary relationships and collective nature of discovery

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Further Viewing

- *The Genius and the Boys* (Bosse Lindquist, Sweden, 2009, color, video, 90 min.). co-produced by BBC, Arte, SVT, NRK and DR. Note: mature content (see supplemental activity).
- *Kuru: The Science and the Sorcery*. (focuses primarily on Michael Alpers) (Siamese Film, Australia, 2010, 52 min.; SBS1365).

Further Reading

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