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TONY ASHBY
Professor Michael Alpers surrounded by the artefacts of his extraordinary career.

The Last Laughing Death

By Jo Chandler
November 13, 2012

After 55 years, the final patrol for cases of the mysterious 'laughing death' in remote Papua New Guinea has returned from the highlands. From this pursuit came Nobel-winning science, clues to 'mad cow' and insights into Alzheimer's disease. It also revealed a little bit of cannibal hidden in us all.

It's 50 years since Michael Alpers, a 28-year-old medical graduate from Adelaide with a restless spirit and an urge "to do health in a different kind of way", hiked into the Papua New Guinea highlands looking for the crucible of a devastating disease epidemic — and stumbled into the crater of an uncharted volcano.

While he smartly sidestepped the sulphuric grumblings of Mount Yelia, young Dr Alpers never really made it back from that trek, succumbing en route to a mystery, a mission, and a culture. This month the now-venerable professor's long expedition reaches its conclusion: The last of the corps of local foot-soldiers he trained over decades to track down and document cases of *kuru* — the name the afflicted Fore people gave to the tremors signalling inevitable and terrible death — are just winding up their final routine surveillance patrols through the villages where the disease once raged.

Today's *kuru* reporters will emerge from their last monthly trek through the mountains and negotiate the rough track north to the provincial capital of Goroka — a four-hour trip, if the route has not washed away in the latest downpour. When they have submitted their final reports to the PNG Institute of Medical Research and collected their last pay cheques, the file will be closed on an epic continuous surveillance effort which began when the first documented reports of the disease emerged in 1957. Along the way, its foot soldiers have navigated some of the most arduous geographical, cultural and humanitarian landscapes imaginable.

Several of the surveyors are second-generation *kuru* sleuths and bush medics, heirs to the stories and skills their fathers acquired in the 1960s when they accompanied Alpers and other pioneering investigators during the height of the *kuru* scourge. Then the mysterious disease was killing up to 200 people a year — mostly women and children — in the Purosa Valley, in the remote Eastern Highlands. It very nearly wiped out the Fore. Locals blamed powerful ritual sorcery for the curse; intrigued medical scientists postulated a genetic cause, or maybe an environmental factor; and patrol officers installed by the Australian administration suspected the Fore tradition of eating their dead — an outlawed practice that had largely ended by 1960. They would all, to varying degrees, turn out to have part of the story.

Fore people recruited to "The *Kuru* Project" worked as translators, guides, cultural advisors, nurses, autopsy assistants, couriers, cooks, security guards, drivers, carriers and custodians of precious human tissue destined for research laboratories in Melbourne, Washington and London. They were instrumental in what is recognised as one of the greatest discoveries in biomedical sciences of the 20th century.

Their involvement was critical to the collection of field data from villages scattered through rugged, remote terrain; the co-ordinated efforts of field workers and scientists ultimately garnered two Nobel prizes (and contributed indirectly to a third). Their continuing surveys have informed and shaped the public-health response to Europe's "mad cow" disease, particularly at its British epicentre, providing warning that a substantial second wave of deaths is inevitable, and that dormant carriers of the infection will long pose a threat to safe blood, organ and tissue supplies. Their legacy also endures in the footnotes of emerging insights into neurodegenerative diseases such as Alzheimer's and Parkinson's diseases.

"We had to climb mountains and cross fast-flowing rivers," one of their original ranks, Taka Gomea, recalled at a Royal Society gathering on *kuru* in London five years ago. "When we approached some villages they tried to chase us away, threatening us with their bows and arrows. We would placate them by giving them salt and other small presents." En route by Dakota aircraft to one patrol site, Gomea recalled an occasion when the cargo doors burst open. He clung to the co-pilot so he could lean out and pull them shut. "I really wanted *kuru* to stop," said Gomea, who eventually became an orderly at a remote medical aid post. "That's why I was happy to work so hard."

His wish was granted. There have been just eight *kuru* cases this century — three in 2000, two in 2001, one each in '03 and '05, and the last in 2009. In each case, it is believed the victim had incubated the disease for an astonishing 50 years or more, having been exposed to infection as a child when participating in mortuary feasts that were an intrinsic part of Fore culture: that is, the cooking and consumption of the dead, every last piece of them, in order to hasten the journey of the departed loved-ones to the land of the ancestors.

Much later, Alpers, who had always felt discomforted by the term cannibalism — "you don't like to call your friends cannibals" — would invent a new term for the Fore ritual: "transumption". It borrowed from the lexicon of Catholic doctrine around the Eucharistic transubstantiation of bread into the body and blood of Christ. He defined the Fore custom as "incorporation of the body of the dead person into the bodies of living relatives, thus helping to free the spirit of the dead". It was a final act of love by the grief-stricken. Yes, as anthropologists had insisted, there was a gastronomic element: people had given ready testimony that humans were delicious, especially their brains. But this was a perk, not a driver, of the practice, Alpers insisted, in papers citing the secrets shared with him and others over decades.

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Despite the deep significance mortuary feasts held for the Fore people and their neighbours, by 1960 the rites had almost entirely ceased, at the insistence of Australian administrators. Immediately, the epidemic began to ease — new cases among children vanished virtually overnight.

Today, the apparent disappearance of the disease means there's little more information likely to be gleaned from *kuru*'s ground zero. As a consequence, this month, the funding that has flowed to Goroka from London since 1996 — when "mad cow" disease (bovine spongiform encephalopathy, or BSE) crossed the species barrier into humans and sent British researchers scurrying to PNG looking for insights from the closely related *kuru* epidemic — finally dries up. And routine field surveillance will come to an end.

There may well be one or two elders who still carry the disease, but there's no expectation that the surveyors' last sweep through *kuru* country will yield another case. Word now travels fast if there's so much as a suspect shiver in the most far-flung village, and it quickly finds its way to the limestone cottage in Fremantle, Western Australia, where Alpers retired after leaving

his post in Goroka as chief of the PNG Institute of Medical Research in 2000.

The walls of Alpers's home are lined with books on PNG and hung with its artifacts and art. Only a couple of years ago he trekked back into the highlands after being summoned to a rumoured case that turned out to be Parkinson's disease. "*Kuru* is still the first thing people think [of] if anyone gets a bit shaky," the professor says. "It's an extremely powerful disease, horrible to live with, and horrible to see someone die of. People are still very much afraid of it."

Alpers remembers the tragedy all too well. In medical literature, the investigation of this "extraordinary disease… will continue to have long-standing significance for neurology, infectious disease and public health", as papers to the landmark Royal Society *kuru* meeting in London in 2008 observed. But for Alpers it is a story populated by individuals with names and faces, children and mothers he tended and held in his arms in the days and weeks before they died, some of whom he cut open within hours of their deaths, searching for the truth of the powerful agent that had claimed them.

The survey teams may have finally left the field, but in Fremantle Alpers continues to sift methodically through his *kuru* archive, looking for new insights. He is still following the trail into the next awakening.

IT WAS WHEN he read the first reports of the mysterious *kuru* in the *Adelaide Advertiser* in 1957 that Michael Alpers — a self-described "disaffected" medical student — became intrigued. A team of local scientists were involved in the early investigation of the disease and Alpers lobbied them to go to PNG, securing a post for himself as a medical officer for the Australian administration which was then considering trying to quarantine the infected region, in a desperate bid to contain the epidemic.

Alpers flew into the wild frontier highlands town of Goroka in 1961. Systematic medical investigation into *kuru* was already well underway, initiated by local district medical officer Dr Vincent Zigas and led by American scientist Dr Carleton Gajdusek, a brilliant, dynamic and controversial figure who would become Alpers's friend and collaborator, and who later won the Nobel prize for his seminal work. A husband-and-wife team of Australian anthropologists, Robert and Shirley Glasse (later Lindenbaum), were also deep in the field, looking for clues in cultural practices and diet.

Alpers spent his first weeks enlisted in a crash course in linguistics that he credits with priming him to deeper engagement with the strange reality he was about to enter. "I learned about languages that had never been written down, about the difference between phonetics and phonemics. It made me realise these languages were extremely complex. Everyone assumed in the outside world — and sometimes still do — that these were primitive people. Well, the languages certainly weren't primitive, and nor was the culture based around the available technology."

He ventured deep into the *kuru* heartland, spending "a couple of months walking around, talking to people". He recalls receiving a particularly warm welcome in a village called Waisa, a solid hour's trek from the nearest road but smack in the heart of epidemic. "People said 'Come, you're very welcome,' and I settled." They built him a hut, which was replaced in time by a house fitted with the luxuries of a water supply and a generator. It would become his home — and later that of his own young family — for long periods. A newer version of it still shelters researchers, and served as the field headquarters for the UK's Medical Research Council Prion Unit.

While *kuru* was Alpers's focus, he could not fail to see the community's other urgent medical needs. "I trained a couple of young

medical clinic.

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> *"It's an extremely powerful disease, horrible to live with, and horrible to see someone die of. People are still very much afraid of it."*

He met Carleton Gajdusek in the field in early 1962. "He'd upset lots of people. Everyone warned me against him, but by then we knew that *kuru* was like scrapie [a transmissible, fatal brain disease which had long occurred in sheep, and was familiar to veterinarians but few others]. Carleton wanted to test the idea that *kuru* was also transmissible, and I did too, so we joined forces. We planned an experiment — collecting autopsy samples [from *kuru* victims], putting them into chimpanzees, and then following them for 10 years." (Alpers stood by — and remains — fiercely defensive of Gajdusek over a murky episode in later years where he was disgraced and jailed for a year for child molestation. He died in 2008.)

The immediate challenge for Alpers was collecting the autopsy tissue from afflicted brains. "When I first got there the local *kiap* [the vernacular for the ranks of Australia's all-powerful field officers — in Okapa] said 'absolutely not, people are fed up [with autopsies], they've disturbed everyone'." Alpers determined much of the distress was due to the bodies being carted to hospital

for examination and then not promptly returned.

His solution was to conduct limited autopsies out in the home villages, assisted by his trained aides and the families of the dead. This had the added benefit of securing fresh tissue very soon after death. He told communities he wanted the brain, nothing else, and that this donation would help scientists find an answer to the deaths. While most villagers were convinced that sorcery was the cause, their experience of the new medical clinics had taught them the concept of "germs" which were treatable. And they were desperate enough to give it a try.

Over the following months Alpers gained autopsy approval from the families of several people dying of *kuru*. Everyone understood too well that no-one recovered from *kuru*, which progressively stole control, mobility, speech but, tragically, not always faculty from the afflicted. Bursting into gales of uncontrollable laughter was another cruel quirk of the disease. "So having established the fact I had permission to do an autopsy I then would go to the *kiap* and he would provide a coroner's certificate — in advance — authorising it," says Alpers. As death approached, Alpers would move into the patient's village, and wait.

It could be a long process. Weeks, usually. "You couldn't do anything else but hang around, a bit like a ghoul. And it normally took a long time even after the patient was moribund, (almost) paralysed, but subsisting on sips of sugar water." Alpers would ready his instruments and prepare a hut for the autopsy, setting out labelled, sterile vials for the tissue he would soon collect.

On the professor's Fremantle kitchen table lies a black and white photograph of the little girl whose story he now recounts, so many years later. She was the second of five field autopsies Alpers conducted during that period. The photo is a still taken from some of the kilometres of footage he filmed over the years, to document the progress of the disease. The girl is leaning on a stick in front of some village huts, looking shyly into the camera. "That's Kigea," he says, tapping the picture. "She was from my own village, Waisa. A wonderful little 11-year-old girl who died of *kuru*." Her end was lingering and awful. Her father despaired and ran away until it was over; her mother had already long-since been claimed by *kuru*, as so many young women were.

"The extraordinary thing was that I could still make contact. I remember asking Kigea the day before she died to put her tongue out. She was in this locked-in state, but she wasn't paralysed, and she understood me talking to her. It was a terrible situation for everybody." Finally, Alpers received the call — she was gone.

After each death, he says, "I would go and talk to the family again, and say, 'Okay?'. They had participated in cutting up bodies in the past — so that was not an unusual activity for them. We had to clear a few people — particularly the women who were wailing. But some of the women stayed. The ones involved put on masks to protect the tissue and I had gloves.

"The father, or a close relative, would hold the head, and I would take the top of the skull off with a bone handsaw. It would take maybe 20 minutes… like cutting an avocado. I would go to particular parts of the brain… take out small cubes. My assistant would hold out the bottle that was relevant, take the lid off, and I'd pop it in.

"Then I'd take the whole brain out and put it in a bucket full of formalin and cotton wool so it wouldn't be deformed, and put the lid on. All our samples would go into an insulated box. Then I put the skull cap back on, and sewed up. Then we said goodbye… gave everyone a hug, and took off. I did this five times. It was enough."

The insulated box would be taken immediately to the station at Okapa and put in the freezer. The next morning a chartered aircraft would come to collect it and fly it to Lae.

From Lae the box would catch a flight direct to Melbourne, where it was collected by a hematologist from the Commonwealth Serum Laboratories, who would store the tissue in a deep freezer until it could be sent to the United States National Institutes of Health in Washington DC where Carleton Gajdusek had assembled some chimpanzees in a primate facility for the next phase of the experiment.

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In early 1964 Alpers followed the samples to Washington, where he would spend the next four years exploring *kuru*'s epidemiology and genetics, but even at that distance, the emotional backwash was inescapable. By now the tissue he had taken from Kigea and another young boy called Eiru had been inoculated into a pair of chimps — Daisey and Georgette — and Alpers would visit them twice a week, examining them, shooting footage, growing fond of them and their quirks and characters. "Chimps are so close to humans it made them difficult to use in lots of ways, but we felt we had to do it."

Two years into the experiment, Daisey and Georgette started behaving strangely. Their gait changed, they had difficulty walking, and lost co-ordination. When they couldn't pick up pieces of apple and put them to their mouths they improvised, using their lips to scoop the apple from the ground. Alpers had seen it all before.

"One day I was examining them and wrote down 'clinical impression — *kuru*'. It was just striking. The tremors, the gait… the intension tremor which is a classic sign of cerebellar disease, which is what *kuru* is." Gajdusek was again in PNG and Alpers sent a telegram summoning him back. By the time he arrived a week later, "Daisey was falling all over the place… it was awful. But at the same time there was this elation that our experiment was going to be successful."

Alpers was convinced Daisey was sick from the agent that had come to her from Kigea, and Georgette from Eiru's brain, but it would be several more months before that could be proved, with samples taken from Georgette's brain at autopsy travelling to London for scrutiny by a neuropathologist.

The day the telegram arrived back in Washington advising that the chimp's brain pathology was "indistinguishable" from human *kuru* "we knew transmission was true". Alpers, Gajdusek and their colleague Joseph Gibbs "wrote our paper in a day, dividing it in three, and posted it at midnight".

Unusually the paper identified Kigea and Eiru — as well as Daisey and Georgette — each by name. Normal scientific convention is to scrub the documentation clean of such emotionally charged contaminants as identity. But in this instance, somewhere in their haste, the scientists' instinct was to give credit where it was painfully due.

The paper appeared in the journal *Nature*, just two weeks later. It was a watershed finding, identifying *kuru* as a new category of infectious disease that caused the degeneration of the brain and nervous system, one that was capable of crossing the species barrier and which passed via unidentified agents lurking within brain matter. These agents would later be identified as a single infectious, self-propagating protein — which broke all previously assumed rules, in that they did not possess nucleic acid. They were given the moniker "prions", and their identification earned another scientist (Stanley B. Prusiner) a Nobel prize. Prions bore the distinction of being the first new pathogen identified in more than a century.

MOMENTOUS AS THE 1966 breakthrough was, *kuru* remained elusive. What was the mechanism spreading the contagion? Carleton Gajdusek resisted what he felt was the too-glib notion that consumption of human flesh was to blame. He argued that the infection might have travelled through cuts or sores or dabbing of eyes during ritual handling of the dead's organs. Local *Kiap* Jack Baker reckoned the scientists were overthinking it, overlooking the obvious.

Alpers had by then spent several years reviewing the epidemiology of the disease, trawling through data collected by patrol officers, scientists and missionaries. Their work had been ably assisted by the Fore people's formidable collective memory — "cause of death is always known, even going back three generations", explains Alpers.

He combined the charts with the insights of anthropologists working in the field, and the secrets of Fore ritual that had been entrusted to him. The Fore's complex eschatology declared that each individual had five souls; that after death they travelled the land on a kind of farewell tour from which ultimately — assuming various rituals over a period of years were honoured — they would be reunited in the land of the ancestors. The most efficient path to this hereafter was for the body to be eaten.

As Alpers, with Jerome Whitfield and other colleagues summarised in a recent paper: "If the body was buried it was eaten by worms; if it was placed on a platform it was eaten by maggots; the Fore believed it was much better that the body was eaten by people who loved the deceased than by worms and insects. By eating their dead, they were able to show their love and express their grief."

It was the women's responsibility to eat the dead, grinding the bones and cooking the flesh, indulging their children along the way with the tastiest bits. Particular body parts were given to particular female kin. Although small boys joined in the feasting, they were generally excluded after about age 10.

By 1964 Alpers had solid figures on *kuru* deaths spread over seven years. "I compared the data for 1957, '58 and '59 with '61, '62 and '63, and looked for any changing patterns. Overall, there was not much, but if you looked at the young kids, the disease had essentially disappeared — even in that short time. This was a major change." Obviously there had been some social or environmental shift. But so much in the Fore world had been in a state of upheaval during that era.

"We made a list, Carleton and I, and there were lots of changes. The introduction of new foods, new animals, the cessation of certain activities. But the one that was biologically the most relevant was the mortuary practices, at least in my view." A couple of years later, field surveys confirmed the disease had died out in children younger than 10 — which fitted with the *kiap*s effectively administering new rules of behaviour through the district. The rules were, says Alpers, "No fighting, build roads, no cannibalism, no child marriage, and plant coffee. And they did it."

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When Alpers put his data together for a presentation in Washington in 1967 "the argument for cannibalism — and I don't use that term anymore, but it was used then — was compelling. Everything fitted. Why did women and children get the disease? Because they were the ones that carried out the practice — the men didn't. It explained why it was dying out in young children — because the *kiaps* had proscribed cannibalism. You could also conclude that the disease was not being transmitted vertically from mother to child. No one born since 1960 was coming down with *kuru*. The penny dropped".

The humbling lesson for scientists and doctors was that while their labours might have helped solve the puzzle, they had not halted the disease. The honour for the life-saving intervention belonged to the officers, both black and white, who administered the new laws of the land.

One aspect of the mystery remained unsolved: where had the disease started? Robert and Shirley Glasse had walked the trail through the signposts of Fore memory to a location and a moment early last century, tracing oral accounts of the disease appearing from the 1910s. In 1970 Alpers wrote a paper proposing that *kuru* had spread from a single case of spontaneous Creutzfeldt-Jakob Disease (CJD) — a rare but real and well documented occurrence in any human population. Ordinarily, such an event might present an individual and a family with an unfathomable tragedy, but it ended there. However, in a culture in which brain tissue was consumed through the Fore's funeral rituals, that single episode amplified over a generation or two, and then took off.

Genetic analysis would later provide evidence supporting this theory. It would also yield a few more surprises: some of these would be critical in managing future disease; others would go back through history, identifying skeletons in the darker recesses of humanity's closet.

When Michael Alpers arrived in New Guinea, in 1961, and visited villages where the women of child-bearing age were all but gone, the view was that the Fore were on the brink of extinction. But what if, in a classic enunciation of Darwinian natural selection, the Fore were merely en route to a population bottleneck, from which the survivors would emerge with more protective genetic pedigrees?

In 2003, Alpers was one of the authors of a sensational paper published in the journal, *Science*. A new round of genetic analysis of elderly survivors of Fore mortuary feasts had revealed they possessed a particular form of a gene which seemed to give them genetic resistance to *kuru*. Wider sampling across 2000 people from other cultures found the same chromosomal quirk. The authors argued that the widespread appearance of this prion-protective gene indicated natural selection had been at work before in this context, and that the consumption of human flesh — and consequent outbreaks of infectious disease — had occurred widely in the remote human past.

WITH *KURU* HAVING ALL BUT VANISHED, and the mechanism for its spread having been eradicated, the whole episode might have quickly disappeared (except in the memories of the depleted Fore) into the annals of curiosity. But then came the "mad cow" crisis. It made international headlines when BSE manifested in British beef in the mid-1980s, as a consequence of beasts having been reared on meal derived from the recycled offal of their own kind. The fear was that consumption of diseased beef would see the disease spread to humans.

That fear was realised when the infection turned up in human form as variant Creutzfeldt-Jakob Disease (vCJD), from 1996. With 176 cases confirmed to date in the UK alone (another 49 are recorded elsewhere, half of them in France), it has not reached the epidemic proportions once anticipated. Part of its management and containment is credited to the early recognition of its similarities to *kuru*. Comparing the medical film archives, the British casualties of vCJD look like pale ghosts of the Fore, their faces contorting into the same anguished shapes, their limbs staggering and trembling in a now familiar dance.

"I spoke to Michael before vCJD appeared, on the basis that we thought it might transmit to humans," recalls neurologist Professor John Collinge, director of the UK Medical Research Council Prion Unit. "Of course it turned out that it did. It seemed to me that *kuru* was the major experience we had of one of these diseases. And we had better get to know as much about it as we can."

Soon Collinge was en route to PNG. His first urgent question of the team on the ground was whether prion disease really could incubate for upwards of 50 years? The findings by his own unit, published in *The Lancet*, would confirm it. "This was quite

extraordinary. It was clear these were long transmissions. And understanding that was then very important in the UK and other countries with vCJD.

"We [also] wanted to see what strain or strains of prions caused *kuru*. We actually found two different strains in *kuru*, which were two of the same strains that cause sporadic CJD which occurs rarely, and at random, in all human populations.

For Collinge, like Alpers, the journey to PNG's *kuru* country took him into uncharted territory. At home he had a still undefined public-health emergency to manage, together with the demands of voracious first-world. How many people had been exposed to killer hamburgers? How many might ultimately be struck down?

In PNG Collinge found himself negotiating more nuanced but no-less confronting questions. What were the obligations of medical investigators, pursuing a quest on behalf of their own populations, to people of the developing world? "We couldn't turn up as westerners and say 'we're only interested in *kuru* — you're dying of malaria, but that is not our interest'. It was ethically essential that we work with the community, contribute... tend to patients with common infectious diseases that might be lethal without treatment, in the same way they were helping us with our medical problem."

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The outcome of this thinking is that an enduring funding and co-operation agreement was forged between London and Goroka. Over the years, the UK project helped build three schools in the *kuru* area, trained teachers and established clean water supplies.

At the heart of the project were the critical clinical lessons to be learned from the Fore. Collinge cited these to the British press, priming his home audience for the likelihood of a second wave of vCJD cases. This warning emerged from genetic analysis of the long-term survivors of *kuru*, who have inherited a profile that acts to delay onset. Scrutiny of *kuru* cases reveals that a key gene shapes the body's defences against the disease, and it exists in two forms: version-m and version-v. People with two identical copies MM or VV (one copy of the gene from each parent) were the first to get *kuru*, while those with one M and one V form (MV) might survive for decades. The same genetic profile is underwriting vCJD, the proportions suggesting another 250 delayed cases may emerge in the UK in the next few decades.

But there is another level of concern in the UK, says Collinge, around what is sometimes described as "modern" cannibalism — the recycling of blood, tissue and organs through medicine. In July this year, while Britain was distracted by the Olympics festival, the UK Health Protection Agency published findings from a survey of appendix tissue removed routinely from Britons in an attempt to measure the prevalence of vCJD across the population. It identified abnormal prions in 16 of 32, 441 cases.

"I think that is quite a worrying figure," Collinge says, "suggesting that one in 2000 people in the UK population are infected. Now these individuals are healthy. Will they ever develop CJD? Are they individuals with very long incubation periods like we see in *kuru*? Some of them possibly. But I suspect a majority of them will be genuine carriers — infected, but they will never develop the disease themselves. They do, though, represent a risk to others if they are blood donors or donate organs. This is an ongoing public health issue in the UK... and *kuru* still has things to say about that."

In the broader medical context, *kuru* continues to provide insight into neurodegenerative diseases such as Alzheimer's and Parkinson's. "It turns out that all these diseases involve accumulation of proteins... They all involve one of the body's own proteins going wrong and forming clumps of material in the brain. It's this process that seems to be critical to what is going on in prion disease [such as *kuru* and vCJD], and it's now becoming increasingly apparent that similar sorts of processes are at work in Alzheimer's and Parkinson's," Collinge says.

IT'S THREE YEARS SINCE the *kuru* pathogen stirred to claim its most recent victim, a 61-year-old woman from a far-flung hamlet right in the heart of the defined disease sector. It had slept within her for over 50 years — suppressed, according to the latest findings — by an accident of chromosomes programmed to resist the infection. She can have been only a young girl when she ate the morsel that would eventually kill her — very likely taking it from the hand of her mother or one of her aunts.

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In the medical literature today *kuru* is recognised not as "an exotic, strange and unique disease caused by cannibalism on a remote island", but as representative of a whole novel class of disease and carrying powerful, enduring lessons for human health.

In PNG's Eastern Highlands, the disease is an unforgotten horror, one many still blame on sorcery, even if they give credence to the interventions and arguments of outsiders who tell them that it was their funeral practice that spread the curse. Through the stories of the *kuru* scouts and the presence of so many outside researchers and doctors over so many years, there is also wide understanding that the Fore's tragedy has provided hard lessons to the wider world.

"In some ways the story has come to its end," Alpers says, even as he knuckles down to a review of every one of 2,700 *kuru* files, a task he estimates will take him two years. There are one or two angles in them he hasn't explored. Meanwhile in laboratories elsewhere, scientists continue to poke and probe secretive prions. "It seems now that every neurodegenerative disease — Alzheimer's, Parkinson's, Huntington's — all have a similar prionic process, though they are not infectious in the same way."

There was supposed to be some fanfare this October, to mark the end of 50-plus years of field surveillance of *kuru*. Alpers and Collinge and others were to return to Goroka. But the plans unravelled after a death in the family of one of the principal players. Instead, a low-key celebration will take place at some point. Meanwhile the project overseers invited the ranks of their retired and serving *kuru* field reporters to nominate what they might like to mark the moment. A medal? A citation?

"They wanted boots," Alpers says, so that is what they got – good, solid walking boots. After all those miles, up and down all those mountains, on the trail of an elusive killer, the *kuru* trackers proudly lace up their hard-earned trophies and continue on their way.

For more information:

'Kuru: the Science and the Sorcery', a documentary by Siamese (/web/20150318111247/http://www.kuru-doco.com/), DVDs available: info@siamese.com.au

'The end of kuru: 50 years of research into an extraordinary disease', 2008 Special Edition, Philosophical Transactions of the Royal Society (/web/20150318111247/http://rstb.royalsocietypublishing.org/site/2008/kuru50.xhtml.), edited by John Collinge and Michael Alpers.

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47 COMMENTS ON THIS STORY

by Roger Colclough

Is there a test for the kuru prion protein ? Obviously this is of some concern to me, my children and the recipient(s) of my having donated bone to one or more recipients. Em tasol, (and hopefully not), pinis.

October 9, 2013 @ 8:39pm

by Dr Momia Teariki-Tautea

I have the fortune of having done research on Childhood diarrhoea / typhoid through the PNGIMR in 1991 and got to know Prof M Alpers, such a humble and knowledgeable man.

January 3, 2014 @ 7:19pm

by Wilson Thompson Orlegge

A remarkable story and a journey that has provided insight into this dreaded disease. Thank you to the writer who wrote in simple terms to the life of Dr Alpers

January 25, 2014 @ 4:10am