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Uncovered brain disease "resistance gene" could offer insights into CJD

19 November 2009

A community in Papua New Guinea that suffered a major epidemic of a CJD-like fatal brain disease called kuru has developed strong genetic resistance to the disease, according to new research by Medical Research Council (MRC) scientists.

Kuru is a fatal prion disease, similar to CJD in humans and BSE in animals, and is geographically unique to an area in Papua New Guinea. In the mid 20th Century, an epidemic of kuru devastated a population in the Eastern Highlands of Papua New Guinea. The infection was passed on at mortuary feasts, where mainly women and children consumed their deceased relatives as a mark of respect and mourning. This practice was banned and ceased in the late 1950s.

Scientists from the MRC Prion Unit, a national centre of excellence in prion diseases, assessed over 3000 people from the affected and surrounding Eastern Highland populations, including 709 who had participated in cannibalistic mortuary feasts, 152 of whom subsequently died of kuru. They discovered a novel and unique variation in the prion protein gene called G127V in people from the Purosa valley region where kuru was most rife.

This gene mutation, which is found nowhere else in the world, seems to offer high or even complete protection against the development of kuru and has become frequent in this area through natural selection over recent history, in direct response to the epidemic. This is thought be perhaps the strongest example yet of recent natural selection in humans.

Lead author Professor John Collinge, Director of the MRC Prion Unit said:

> *"It's absolutely fascinating to see Darwinian principles at work here. This community of people has developed their own biologically unique response to a truly terrible epidemic. The fact that this genetic evolution has happened in a matter of decades is remarkable. Kuru comes from the same disease family as CJD so the discovery of this powerful resistance factor opens up new areas for research taking us closer to understanding, treating and hopefully preventing a range of prion diseases."*

The study "A Novel Protective Prion Protein Variant that Colocalizes with Kuru Exposure", which began in 1996, will be published in the New England Journal of Medicine on 19 November 2009.

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Notes to editors

1. Study available on request.

2. For almost 100 years the Medical Research Council has improved the health of people in the UK and around the world by supporting the highest quality science. The MRC invests in world-class scientists. It has produced 29 Nobel Prize winners and sustains a flourishing environment for internationally recognised research. The MRC focuses on making an impact and provides the financial muscle and scientific expertise behind

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medical breakthroughs, including the first antibiotic penicillin, the structure of DNA and the lethal link between smoking and cancer. Today MRC funded scientists tackle research into the major health challenges of the 21st century. www.mrc.ac.uk

3. Prion diseases or transmissible spongiform encephalopathies (TSEs) belong to group of progressive conditions that affect the nervous system in humans and animals. In humans, prion diseases impair brain function, causing memory changes, personality changes, a decline in intellectual function (dementia), and problems with movement that worsen over time. They are fatal conditions. Familial prion diseases of humans include classic Creutzfeldt-Jakob disease (CJD), Gerstmann-Sträussler-Scheinker syndrome (GSS) fatal insomnia (FI) and Kuru.

4. Kuru was restricted to the Fore linguistic groups and their immediate neighbours which whom they intermarried. It was the practice in the Fore society for kinship groups to consume deceased relatives at mortuary feasts, a practice that resulted in human-to-human prion transmission. On the whole, men and children over 8 years of age did not participate in the feast, with the result that kuru at its peak predominantly affected women and children. As recorded in oral history the first cases appeared in the early 20th century and thereafter the number of cases increased in incidence. A peak annual mortality of more than 2% was recorded in some villages. Some villages became largely devoid of young women. More information on the Papua New Guinea Institute of Medical Research is available here www.pngimr.org.pg.

5. The study was lead by the Medical Research Council Prion Unit, and involved scientists from the University College London Institute of Neurology; Papua New Guinea Institute of Medical Research; and Curtin University Australia. Other institutions participating; the Genome Centre, Barts the London Queen Mary's School of Medicine and Dentistry; London School of Hygiene and Tropical Medicine.

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