

Clinical Research Studies in the UK and PNG

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Title of project or programme

Clinical Research Studies in the UK and PNG

Principal Investigators of project/programme grant

Title	Forname	Surname	Institution	Country
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Address of institution of lead PI

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- United Kingdom

Source of funding information

Medical Research Council

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2790679.21

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Total duration of award in months

60

The project/programme is most relevant to

- Prion disease

Keywords

Research abstract in English

Kuru is one of a closely related group of neurodegenerative conditions which affect both humans and animals known as the transmissible spongiform encephalopathies or prion diseases. Animal prion diseases include scrapie, a naturally occurring disease affecting sheep and goats, which has been recognised for over 200 years and is present in many countries world-wide, and the much more

recently recognised bovine spongiform encephalopathy (BSE) amongst cattle. The human prion diseases have been traditionally classified into Creutzfeldt-Jakob disease (CJD), Gerstmann-Straussler-Sheinker syndrome (GSS) and kuru.

Kuru provides the principal experience of an epidemic human prion disease. It has steadily declined in incidence following the abrupt cessation of its route of transmission, endocannibalism, in Papua New Guinea in the 1950s. The arrival of the novel acquired prion disease, variant CJD, and the unknown prevalence of infection following the extensive dietary exposure to BSE prions in the UK, has led to renewed interest in kuru, particularly with respect to the range of possible incubation periods and the effect of genetic susceptibility factors.

That the incubation period of infection acquired by dietary exposure to human prions can exceed 50 years, as found in kuru, suggests the need for caution with respect to predictions of vCJD epidemic size and duration based on the numbers of clinical cases seen to date. Infection of humans with cattle BSE prions involves cross-species, rather than within-species transmission, which would further increase the mean and range of incubation periods.

The research activities have seven major aims.

(1) Identify and study all remaining kuru patients: document the maximum incubation periods: We have studied the clinical features of recent kuru patients and compared clinical and other diagnostic features with other human prion diseases, notably iatrogenic and variant CJD. Surveillance for new patients is ongoing.

(2) Epidemiological analysis of kuru records: Data will be analysed for standard epidemiological variables and differences in geographical area.

(3) Document mortuary feast practices and traditional beliefs of the aetiology of kuru by interview of surviving participants and other members of the Fore community: A large number of interviews have been conducted and are being analysed. More interviews are planned to expand the cultural and geographical range of the study and to probe deeper into the details of the mortuary rites.

(4) To investigate for any evidence of maternal or other routes of kuru transmission: From the data available we conclude that maternal transmission was not a feature of kuru. The data on mortuary practices do not support a high probability of parenteral transmission. Thus the common route of transmission was undoubtedly oral.

(5) Genetic susceptibility: details of laboratory studies are given under Unit Human Molecular Genetics research programme.

(6) Peripheral pathogenesis, tissue distribution and investigation of possibility of asymptomatic carriers of kuru: Tissues obtained from a kuru autopsy are under intense investigation. We will interview elderly survivors of the kuru epidemic in all affected linguistic groups to look at factors affecting the transmission of kuru. We aim to obtain autopsies when these older people die to determine whether any were asymptomatic carriers of kuru.

(7) Data archive and bibliography: A full inventory will be made of all the kuru and kuru-related files. When the appropriate equipment is available archival and contemporary footage will be combined and edited on digital format for making teaching films on kuru, of varying length and complexity suitable for medical educational purposes.

Lay Summary

Kuru is one of a closely related group of brain diseases which affect both humans and animals known as the transmissible spongiform encephalopathies or prion diseases. Animal prion diseases include scrapie, a naturally occurring disease affecting sheep and goats, which has been recognised for over 200 years and is present in many countries world-wide, and the much more recently recognised bovine spongiform encephalopathy (BSE) ("mad cow disease") amongst cattle. The human prion diseases include Creutzfeldt-Jakob disease (CJD) and kuru.

Kuru provides the principal experience of an epidemic human prion disease. It has steadily declined in incidence following the abrupt cessation of its route of transmission, endocannibalism, in Papua New Guinea in the 1950s. BSE transmitted to humans causes variant CJD. The unknown prevalence of infection following the extensive dietary exposure to BSE prions in the UK has led to renewed interest in kuru, particularly with respect to the range of possible incubation periods and the effect of genetic susceptibility factors.

Present studies are ongoing to record the maximum incubation period for kuru. Epidemiological studies that include genetic and anthropological data are being conducted to help us fully understand the kuru epidemic and its implications for the variant Creutzfeldt-Jakob disease epidemic.