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Variant Creutzfeldt-Jakob disease: prion protein genotype analysis of positive appendix tissue samples from a retrospective prevalence study

A preliminary research finding casts doubt on the current estimates of vCJD incidence. They could be underestimates.

The study examined tissues from 12,674 samples retained after surgery. Samples from three people were found to have evidence of prion disease. Two of these samples were tested for genotype at the codon 129. All clinical cases of vCJD to date have been of the type MM which forms a substantial minority of the population.

Both samples were found to be of the type VV (the remaining possibility being type MV). This is the first time there has been any suggestion that the VV genotype could be susceptible to vCJD.

Comment

The emergence of a new genotype associated with vCJD opens up the possibility that different genotypes have different latency periods. If the VV genotype has a long latency, including a long period where there are low levels of disease agent in body tissues, then blood transfusions etc. could perpetuate the disease in the human population long after cases caused through oral exposure have ceased to manifest.

Recent studies of kuru have shown that this transmissible prion disease does affect the MV genotype and that this genotype experiences longer incubation times. [*Lancet* 2006 Vol.367 p 2068 – 2074].