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## Fears raised over vCJD infection

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An unusual case of variant CJD, the human form of "mad cow disease", has resurrected fears that some people might still be infected and yet to develop symptoms.

The genetic make-up of a 30-year-old man who died from vCJD suggests some cases of infection might have very long incubation periods, possibly spanning decades.

If this is true there could be an unknown number of infected people alive who are unaware they have the condition.

They may have acquired the disease from eating beef contaminated with the cattle disease bovine spongiform encephalopathy (BSE) as young children. Whether or not they develop the terrible symptoms of vCJD and die from the condition would depend on how long it takes to develop.

Many infected individuals could die from other causes before reaching the end of the incubation period, experts believe.

Fears of a major epidemic claiming thousands of lives surfaced at the height of the "mad cow disease" scare in the 1990s. The disaster never materialised, and since 1994 only around 200 cases of vCJD have been confirmed worldwide.

In the UK the total number of definite and probable cases of vCJD now stands at 170, including four suspected victims who are still alive. To date, every person confirmed as having vCJD has had a particular "homozygous" form of the human prion protein gene known as MM. Rogue prion proteins are believed to be the infective agents behind the disease.

The same gene can also take the form VV or MV. The newly identified victim, who died in January after suffering symptoms of personality change, memory loss, unsteadiness and visual hallucinations, had the "heterozygous" MV form. Scientists investigating the case led by Professor John Collinge, from the Medical Research Council Prion Unit in London, pointed out that related brain diseases had longer incubation periods in people with the heterozygous MV prion gene. They included kuru, thought to be linked to cannibalism in Papua New Guinea, and CJD triggered by treatment with growth hormones.

Writing in The Lancet medical journal, the researchers said some MV patients with kuru had incubated the disease for more than 50 years. The scientists suspect the MV gene might cause similar long incubation periods in people infected with vCJD.

About a third of the general population has the susceptible MM form of the prion gene, which produces a double dose of the amino acid methionine. The researchers wrote: "If individuals with other genotypes are similarly susceptible to developing prion disease after BSE prion exposure, but with longer incubation periods, further cases, which may or may not meet diagnostic criteria for vCJD, would be expected."

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