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## General Neurology

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Dec. 17, 2009  
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### Program Overview

### Learning Objectives

Upon successful completion of this educational program, the reader should be able to:

1. Discuss the results of this study
2. Review the relevance and significance of the study in the broader context of clinical care

### Disclosures

**John Gever, MD; Associate Clinical Professor of Medicine, Harvard Medical School, Boston,** has disclosed that they have no relevant financial relationships or conflicts of interest with commercial interests related directly or indirectly to this educational activity.

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## There May Be More vCJD Risks than Expected

By John Gever, Senior Editor, MedPage Today  
Published: December 17, 2009

Reviewed by **Dori F. Zaleznik, MD**; Associate Clinical Professor of Medicine, Harvard Medical School, Boston and Dorothy Caputo, MA, RN, BC-ADM, CDE, Nurse Planner

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Some people thought to be genetically resistant to variant Creutzfeldt-Jakob disease (vCJD) may in fact be vulnerable to the fatal syndrome, researchers said.

A 30-year-old British man who was heterozygous for a polymorphism within codon 129 of the prion protein gene, *PRNP*, developed the condition and died earlier this year, according to John Collinge, MD, of University College London in England, and colleagues reporting in the Dec. 19/26 issue of *The Lancet*.

Everyone else ever diagnosed with vCJD has been homozygous for a methionine residue at that location, the researchers said.

Scientists believed that individuals who were homozygous for valine or heterozygous would either not contract vCJD or would show very long incubation periods.

Methionine-homozygous individuals make up about one-third of the population in Great Britain, the researchers said.

The disease occurs when a misfolded version of the human prion protein comes into contact with normal versions and catalyzes their refolding into the abnormal configuration.

In this form, the protein causes brain cells to wither away and die, leaving macroscopic holes behind -- hence the name "spongiform encephalopathies," for the family of related conditions that affect a number of species, including cattle, sheep, elk, and deer, as well as humans.

Misfolded prion proteins are pathogenic and transmissible across species, and disease can spread simply by eating tissues containing the proteins.

Variant CJD has been a special concern in Great Britain, where cattle with bovine spongiform encephalopathy entered the human food chain. Since 1994, some 200 cases have been diagnosed worldwide (mainly in Britain) as a result of exposure to prions in beef.

However, other types of acquired spongiform encephalopathies have been known to affect people of all codon 129 genotypes.

Collinge and colleagues cited one case involving the recipient of a blood transfusion from a donor subsequently identified as having vCJD.

The recipient had died of unrelated causes but an autopsy revealed brain lesions consistent with prion disease. He was heterozygous at codon 129.

### Action Points

- Explain that "mad cow disease" in humans is extremely rare, even in Britain where the most cases have been reported. Also explain that most people exposed to prions from diseased cattle or other animals do not develop the disease.
- Explain that a similar and also very rare condition called sporadic CJD happens at random and cannot be prevented or predicted. CJD in any form is untreatable and always fatal, although the time between exposure to the causative agent and development of symptoms can vary.

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It was not known how the man described in the current *Lancet* report contracted vCJD. He had not received any blood transfusions or other human tissue implants, and prion-contaminated beef was thought to have been eliminated from human foods many years ago.

His death came about 20 months after the onset of neuropsychiatric symptoms, including personality change, intellectual decline, and loss of motor control.

No autopsy was performed, but the man's young age, clinical features, and MRI and electroencephalography findings led to the diagnosis of vCJD. This combination of features made sporadic CJD "unlikely," Collinge and colleagues wrote.

"The majority of the U.K. population have potentially been exposed to BSE prions but the extent of clinically silent infection remains unclear," they said in the report.

If the two-thirds of Britons not homozygous for methionine at codon 129 turn out to be susceptible to vCJD but with longer-than-expected incubation periods, additional cases "would be expected," the researchers wrote.

But they added that other genetic factors also affect susceptibility and incubation periods.

"Cases of vCJD to date may have unusual combinations of genotypes at these loci, yet to be fully characterized," the researchers cautioned.

No external funding for the study was reported.

Collinge reported a relationship with D-Gen Ltd., a company involved with prion protein diagnostics, therapy, and decontamination. Other authors reported no potential conflicts.

**Primary source:** The Lancet

Source reference:

Kaski D, et al "Variant CJD in an individual heterozygous for PRNP codon 129" *Lancet* 2009; 374: 2128.

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