FactSheet

Para Su Información

Creutzfeldt-Jakob Disease (CJD)

What is Creutzfeldt-Jakob disease (CJD)?

CJD belongs to a group of neurological diseases known as transmissible spongiform encephalopathies (TSEs). TSE diseases in humans are very rare but fatal. CJD is a slow degenerative human disease of the central nervous system. It is classified as a transmissible spongiform encephalopathy because of the characteristic spongy degeneration of the brain that occurs as the disease progresses. CJD occurs sporadically worldwide at a rate of 1 case per 1 million people per year.

What is variant Creutzfeldt-Jakob disease (vCJD)?

Variant Creutzfeldt-Jakob disease (vCJD) is another rare and fatal human neurologic disease that falls into the category of transmissible spongiform encephalopathies (TSEs). Like Creutzfeldt-Jakob disease (CJD), vCJD causes a spongy degeneration of the brain. vCJD is a new disease, which was first described in March 1996.

How does vCJD differ from CJD?

In contrast to the classic form of , the new variant or variant form (vCJD) in the United Kingdom and France affects younger persons (average age at onset: 28 years), and has different clinical features from CJD. People with vCJD begin with serious psychiatric problems or problems with their senses (ears, eyes or smell), this first set of symptoms is followed weeks or months late by poor muscle coordination, problems with muscle spasms, and mental confusion; these patients also have abnormal electroencephalograms (EEG). The illness lasts for at least 6months, but most people die approximately 13 months after their symptoms begin. When patients' brains are examined by autopsy, there are clear changes in brain tissue structure including many "spongiform" or open spongy-looking areas, abnormal spots of prion protein called plaques, and other areas with much prion protein accumulation.

Exactly how does this newly recognized variant of CJD differ from classical CJD?

In 1996 the Spongiform Encephalopathy Advisory Committee (SEAC) or the UK announced the identification of 10 cases of variant CJD (vCJD, Lancet, 1996, 347:921-25). The following features describe how vCJD cases differ from the sporadic or classical form of CJD:

- The affected individuals were much younger than the classical CJD patient. Typically, CJD patients are over 63 years old. The average patient with vCJD is 28 years old; patients ranged from 12-52 years old.
- The course of vCJD averaged 13 months. Classical CJD cases average a 6 month duration.
- In the vCJD cases, electroencephalographic (EEG) electrical activity in the brain was not typical of classical CJD.
- Although changes in brain tissue structure of patients with vCJD were recognizable as CJD , the pattern was different from classical CJD, with large aggregates of prion protein plaques often surrounded by vacuoles.

How did people get this new variant of CJD?

On March 20, 1996 a statement from the Spongiform Encephalopathy Advisory Committee (SEAC) of the United Kingdom indicated concern that before November 1989, when inclusion of certain cow and sheep by-products in human food was banned, the BSE agent may have been transmitted to people

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through contaminated food products. The SEAC said that food might account for the 10 vCJD cases described in April, 1996 in the medical literature (Lancet 1996;347:921-5). The specific foods, if any, that may be associated with the transmission of this agent from cattle to humans are unknown. However, the SEAC has indicated that milk and milk products are unlikely to pose any risk for human exposure to the BSE agent.

What is the evidence directly linking this newly recognized variant of CJD to BSE exposure?

There is strong epidemiologic and laboratory evidence suggesting that new variant CJD (vCJD) and BSE are caused by the same infectious agent. For instance, there have been no confirmed cases of vCJD in other geographic areas where there have been no BSE cases. In addition, the time interval or "incubation period" between the most likely period for the initial exposure of the population to potentially BSE-contaminated food (1984-1986) and onset of initial vCJD cases (1994-1996), about 10 years, is similar to the known time intervals between exposure to the classical CJD agent and the development of CJD.

An experimental study reported in June 1996 showed that three cynmologus macaque monkeys that were injected with brain tissue from cattle with BSE later developed symptoms and changes in brain tissue that were strikingly similar to vCJD (Nature 1996; 381:743-4). Another study published in 1996 showed that prions obtained from 10 vCJD patients and BSE-infected animals had molecular characteristics that were similar to each other but that distinct from prions obtained from patients with classical CJD (Nature 1996;383:685-90). Furthermore, intermediate results of an ongoing experimental study involving injection of a panel of mice (Nature 1997;389:498-501). A recent study using transgenic mice (PNAS 1999;96:15137-15242) supports the hypothesis that the BSE agent from cattle causes vCJD.

How many cases of variant CJD have occurred?

Cases of variant CJD are very rare, and most have occurred in the United Kingdom. The latest information (October 2, 2000) issued by the Department of Health, United Kingdom (www.dohh.govv.uk/cjd/) indicates that there have been 73 confirmed cases of vCJD in the United Kingdom. These cases have all been diagnosed since 1995. France has reported two cases. The Republic of Ireland reported one case in 1999. No cases have been recognized in other European countries, or in the United States.

Could anyone in Europe diagnosed with the newly recognized variant of CJD (vCJD) have acquired this from vaccines?

No evidence exists that any case of vCJD has resulted from administration of a vaccine, and no cases of vCJD has been reported in the U.S. In the UK, the majority of cases of vCJD were born before 1980, and it is very unlikely that they received vaccines contaminated with the BSE agent (Vaccine 2000 19;409-410). Surveillance of vCJD in the UK has identified three "risk factors," or characteristics common to most if not all of the people who had had vCJD: 1) residence in the UK; 2) a particular genetic susceptibility; and 3) age. Epidemiological evidence to date suggests that these cases of vCJD acquired the disease from eating beef products containing the BSE agent after 1980. To date (October 2000) there have been 76 confirmed cased of vCJD. Of these, 73 have occurred in the UK. A case of vCJD was reported in the Republic of Ireland in June 1999, in a person who had been a UK resident from 1989 to 1995. Two cases of vCJD have been diagnosed in France, one in 1996 and the other in 1999. Neither of these people had lived or traveled in the UK. However, according to data published by the UK (HM Customs and Excise), France was a leading European importer of bovine products during the period 1980-1996. No other cases of bovine products during the period 1980-1996.

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