

vCJD Questions & Answers

What is vCJD?

Variant Creutzfeldt-Jakob disease (vCJD) is a rare, incurable, fatal neurodegenerative condition. It is a prion disease that was first described in 1996 in the United Kingdom. There is strong scientific evidence that the agent responsible for the outbreak of prion disease in cows, bovine spongiform encephalopathy (BSE or 'mad cow' disease), is the same agent responsible for the outbreak of vCJD in humans.

How is vCJD Diagnosed?

vCJD has unusually long incubation periods measured in years. Currently, a diagnosis can only be confirmed following examination of the brain after death.

Blood donation restrictions in Bermuda are based on how long you spent in the UK or Europe. How were these set?

BHB follows guidelines used for blood donation in the US. Like the US, Bermuda residents did not have a wide-spread exposure to UK meat products.

People who spent more than three months in the UK between 1980 and 1996, when infected meat products were sold, cannot donate. After 1996, the UK instituted and enforced rules to prevent contaminated cattle from entering the human food chain, which greatly reduced exposure, and so after 1996 there is no restriction on donating blood in Bermuda.

The recommendation for the maximum time spent in Europe is five years. The longer time period reflects that the risk of exposure was much lower. The French imported at least 5% of their beef supply from the UK before 1996.



What are the symptoms of vCJD?

The clinical picture often starts with psychiatric symptoms, such as anxiety and depression. There may be persistent pain, with odd sensations in the face and limbs. These symptoms are followed by more obvious neurological symptoms and progressive dementia. People with vCJD tend to live longer than people with most other forms of CJD, with an average of 14 months (six to 114 months) between symptoms starting and death.

How long does the onset of symptoms take and would it be possible to institute a definitive deferral period instead of an indefinite one?

The incubation period of prion diseases such as vCJD can span several decades. Individuals infected in the 1990s might not yet have developed symptoms. We simply do not know, at present, how many people have been exposed to prions, and what the implications of this might be for the blood donor pool. As such, it would be impossible to state a definitive deferral period for those at high risk of infection.

Are there any blood tests for vCJD?

Blood tests are used worldwide to prevent transfusion-transmitted infections of known pathogens. Unfortunately, there is currently no suitable test for vCJD. The types of tests that are used to screen blood cannot be applied to vCJD because of the different type of infectious agent (abnormal prion protein rather than bacteria or virus).

There is some hope. Researchers in the UK have developed a blood test which seems to be sensitive enough to detect abnormal prion in the blood of some patients with clinical vCJD. However, it is uncertain whether the test will prove sensitive enough to detect infection during the incubation period where there are no symptoms of infection.



Can BHB review the restrictions regarding blood donations for people who have lived in the UK and Europe between 1980 and 1996?

International organisations took the step of limiting people who had lived in the UK between 1980 and 1996 to assure safety of the blood supply by reducing the risk from vCJD. People who have been in the UK during this time may have been exposed to the vCJD agent, which is believed to be the same as the agent of bovine spongiform encephalopathy (BSE, or 'mad cow' disease).



It is unlikely that this restriction will change at this stage. The ongoing risk posed by vCJD is still being examined at international organisations. Available evidence from leading experts suggests that the risk of secondary transmission of vCJD through blood transfusion remains "significant".

Much is still not understood about vCJD and there is no data to support it being dismissed as a threat. Though cases of vCJD are now rare, given the level of uncertainty regarding the potential for blood borne transmission, precaution remains the guiding principle in decision making.

Recent studies indicate that tens of thousands of people in the UK might be 'silent' carriers of the prions responsible for the disease. This raises the possibility that hundreds of blood donors could transmit the infection on to others through the blood supply.

Official statistics state that 229 people worldwide – 177 in the UK (where the BSE crisis primarily took place), 27 cases in France and 25 cases elsewhere in the world – had definite and probable disease since it was first identified nearly 20 years ago. Only one case has been seen since 2012. In December 2003, the first presumed transmission of vCJD by blood transfusion was described. The transfusion occurred in 1996; the blood donor was well at that time but went on to develop symptoms of vCJD in 1999. The recipient was diagnosed with vCJD in 2003.

Since then, two other cases associated with blood transfusions have been identified. With an incubation

period that could stretch decades, it is hard to say that we have seen the end of the outbreak and that it is over. If there are people who are carrying the agent of vCJD, it may be several years before we can predict their fate.

Unfortunately, there hasn't been a breakthrough in the development of a specific screening test for vCJD. Creating a reliable way to test blood for the prions linked to vCJD would be a major achievement, but it remains a technical scientific challenge.



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