



IMMUNE DEFICIENCY FOUNDATION

The National Organization Dedicated to Research, Education and Advocacy for the Primary Immune Deficiency Diseases.

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FREQUENTLY ASKED QUESTIONS ABOUT vCJD
AND IMMUNE GLOBULIN

1) What is vCJD?

Variant Creutzfeldt-Jakob Disease (vCJD) is a rare and fatal disease that affects the structure and function of the brain. vCJD is related to Mad Cow Disease that affects cattle.

2) What causes vCJD?

vCJD is thought to be due to prion proteins, a modified form of a normal cell protein.

3) What are the signs and symptoms of vCJD?

vCJD generally affects younger persons, average age 26 years. The initial symptoms are often serious psychiatric problems or problems with hearing, vision, or smell. After weeks or months, poor muscle coordination, muscle spasms, and mental confusion develop. Individuals are generally sick with the disease for one year before dying of complications.

4) Who is at greatest risk for developing vCJD?

People living in the United Kingdom are at greatest risk from eating vCJD infected beef prior to the food bans in 1996. These bans were designed to prevent infected cattle from entering the human food supply.

5) Is vCJD contagious from person to person?

No, vCJD is not contagious from person to person by coughing or sneezing or contact. The only way it could be spread from person to person is by blood transfusion, organ or tissue transplantation, or less than vigorous sterilization of multiple use surgical instruments.

6) How many people have been diagnosed with vCJD?

To date, there are about 150 people who have contracted vCJD in the UK. Some statistical estimates indicate that the number of new cases is declining.

7) In what countries has vCJD been reported?

United Kingdom, France, Republic of Ireland, Italy, Canada, and the United States have all reported cases. The one US case occurred in a long-term prior resident of the UK who moved to the US. 97% of cases have been reported from the United Kingdom. All cases are presumed to be the result of consumption of British beef except for two recent transfusion (blood, not IGIV or other plasma derivatives) cases in the UK.

8) If I travel to the U.K. (or other countries named above), am I at increased risk of developing vCJD?

No, you are not. The risk is linked to eating infected beef. National laws require that such beef be removed from the market.

9) What is the risk of someone contracting vCJD from current US plasma products?

There is no known risk of contracting vCJD from the current US plasma supply, as there are no vCJD cases in the US. In addition, there are donor deferral measures in place to prevent individuals who lived in the UK from 1980 - 1996 or lived in Europe on US military bases that received British beef during the same era from donating blood or plasma.

10) What is the likelihood of someone contracting vCJD from U.S. plasma products that they used before 1999?

The risk is also thought to be zero. The only greater potential risk would be if a US citizen were treated with a plasma derivative, such as IGIV that was manufactured in the UK from plasma collected from UK citizens. A corporation, BPL, was a major manufacturer of these products for the UK in the late 1990's.

11) Have there been any reported cases of individuals contracting vCJD from plasma products?

No, the risk is theoretical, even in the UK. However, some statistical models predict that individuals treated with many lots of plasma products from the UK in the late 1990's would have an increased risk of exposure to the prion proteins.

12) Was plasma from U.K donors ever used in the manufacturing of licensed U.S. plasma products?

No. US plasma products have only been manufactured from plasma from qualified US donors. No UK IGIV products have ever been licensed for sale or distribution in the US.

13) Are any measures currently in place to detect the organism that causes vCJD in plasma products and if so, what are they?

Currently there is no test sensitive or rapid enough to be useful in screening plasma donations. However, some manufacturers have carefully tested their manufacturing processes to determine if they have steps that are effective in removing prion proteins if they were present. ZLB Behring has recently received approval from the US FDA to make a statement about their process. Other manufacturers are similarly testing their processes and some have published their results in the scientific literature. For IGIV production, all manufacturers have some steps that appear to be effective in removing prion proteins if they were present in plasma pools used for manufacturing.

14) Are there any laboratory or other medical tests that I can take to determine if I have been exposed to this organism?

There are no tests available at this time to determine exposure to prion proteins.

15) What type of physician evaluates someone at risk of vCJD?

Your treating physician may refer you to a Neurologist for further evaluation if indicated.

16) Are there any precautionary steps that I can take to reduce my risk of potentially contracting vCJD?

The two risks are:

1. Eating infected beef.
2. Being transfused with blood and theoretically being infused with plasma derivatives like IGIV from a donor with pre-symptomatic vCJD.

Government agencies and IGIV manufacturers have taken steps to address these two risks.

USDA has started a major screening program for US cattle to confirm that US herds are NOT affected by Mad Cow Disease (the cattle form of vCJD). This program will reassure us that there are no infected cattle in the US.

IGIV products licensed and distributed in the US are manufactured from qualified US plasma donors who have never been reported to have vCJD.

17) Are there any steps I can take to stay informed about the safety of the IGIV products in the United States:

Yes, personal record keeping is very important. Each time you or your child receives an infusion of IGIV, the following information should be recorded:

- a. Date
- b. Brand name of IGIV
- c. Lot number of IGIV
- d. Amount of product infused
- e. Any side effects or adverse reactions experienced

Additionally, you should enroll in the Patient Notification System (www.patientnotificationsystem.org or 1-800-update-U) to receive direct notification of any IGIV recalls or withdrawals from the market. The Patient Notification System is a free, confidential, 24-hour communication system providing information on plasma derived product withdrawals and recalls.