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variant Creutzfeldt-Jakob disease (vCJD)

Signs of variant Creutzfeldt-Jakob disease in a patient with hemophilia: FVIII concentrates most likely cause

February 16, 2009 - Health authorities in the United Kingdom have announced that a man with hemophilia was found at post mortem examination to have evidence of infection with the agent causing variant Creutzfeldt-Jakob disease (vCJD). The man died of unrelated causes and never showed symptoms of vCJD. As part of an ongoing study, the patient's spleen was biopsied after his death and evidence of the infectious agent, abnormal prion proteins, was detected. This is the first time that a person with hemophilia has been found to have any evidence of vCJD infection.

In documents distributed on Monday to hemophilia treaters, the UK Health Protection Agency is stressing that, "This new information does not change the way any patients with bleeding disorders will be treated." Although this case suggests that those patients exposed to contaminated products in the past may be at risk for vCJD, it does not mean that current plasma-derived products on the market today carry such risk.

Bleeding is still the leading cause of death in people with severe hemophilia, and it is essential to keep these risks in perspective. The WFH continues to view plasma-derived products as an important treatment option for the global bleeding disorders community. In fact, globally, no clotting factor concentrates manufactured today contain UK plasma. In addition, plasma donors who have spent time in the UK are deferred from donating blood or plasma in their own countries. We will continue to engage with health authorities and other stakeholders to remain vigilant concerning the safety of treatment products.

Background

This patient had severe hemophilia A, died in his early seventies, and eleven years ago he infused clotting factor concentrate which was later identified as having been contaminated with plasma from a donor who went on to develop vCJD after donation. In 2004, patients in the UK were notified about the potential risk from UK-sourced products infused between 1980 and 2000 and so may be at elevated risk for vCJD compared to the general population. ([WFH TSE Task Force Bulletins on vCJD – 2004](#)) The discovery of abnormal prion proteins in the spleen of this man makes this case similar to the cases of transmission of

vCJD through blood transfusion that have been identified in the UK over the past few years. ([WFH Statement on Second vCJD Transfusion-Transmission Case](#))

The patient was a meat-eater, underwent surgical procedures and had received red cell transfusions – all of which are known risk factors for vCJD transmission. However, at this time, investigators believe that the most likely cause of his vCJD infection was the contaminated UK-sourced clotting factor concentrate. A complete medical history is essential for evaluating these risk factors and investigators are still working to complete that part of their study. The judgment that the infection was due to clotting factor concentrates is not final and the WFH will continue to follow this case closely, releasing more information as it becomes available.

Clotting factor concentrates made in the UK from donors who later developed vCJD were exported to several countries in the period 1980-1998. These countries include Brazil, Turkey, Brunei, UAE, India, Jordan, Oman and Singapore. Health authorities in these countries were notified in 2004. In addition, concentrates made from the plasma of UK donors who were never associated with vCJD were exported to an even larger number of countries. The risk from these products is considered very low, but not zero. However, it now seems likely that plasma-derived factor concentrates can transmit vCJD. It is important to remember that this man did not develop vCJD and died from an unrelated cause. It is also important to note that thousands of people with hemophilia in the UK have received factor concentrate that was derived from donors who later developed vCJD. None of these have developed vCJD.

More information will be released in the next few days by the [UK Health Protection Agency](#) and the [UK Haemophilia Centre Doctors' Organisation](#). Check this site for links to the latest updates.