Canadian Hemophilia Society
Help Stop the Bleeding

AHCDC

FOR IMMEDIATE RELEASE

vCJD infection reported in a person with hemophilia in UK: FVIII concentrates considered most likely cause

February 19, 2009 - On February 17, the Health Protection Agency in the United Kingdom (UK) reported 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.

vCJD is the human disease caused by infection with an abnormal prion protein. The principal cause is the ingestion of beef infected with Bovine Spongiform Encephalopathy (BSE) or Mad Cow Disease. In the 1980s and 1990s, hundreds of thousands of cattle in the UK died from BSE. The epidemic of BSE affected Europe to a lesser extent. Fewer than 20 cattle have been diagnosed with BSE in North America. No North American has been diagnosed with vCJD from eating North American beef.

In a February 16 statement, after consulting experts in the UK, the World Federation of Hemophilia (WFH) stated,

> "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... 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."

See www.wfh.org for the complete statement and updates.

Approximately 200 cases of vCJD have occurred in the world since the mid-1990s, principally in the UK. However, only four cases are blamed on the transfusion of red blood cells from a donor who later developed vCJD. If

confirmed, this new case would be the first case of vCJD caused by the infusion of fractioned plasma products such as clotting factor concentrates.

SITUATION IN CANADA

According to manufacturing and medical records, no Canadian has received clotting factor concentrates made with plasma from a donor known to be infected with vCJD.

Approximately 30 Canadians, however, may have used factor XI (factor eleven) concentrates to treat factor XI deficiency made from UK-source plasma in the period 1992-1998. In 2004 Canadian health authorities worked with physicians to notify these patients of the estimated low risk of contracting vCJD from these products.

The Association of Hemophilia Clinic Directors of Canada and the Canadian Hemophilia Society are recommending that physicians notify these 30 patients of this latest development.

The AHCDC and CHS believe that clotting factor concentrates being used in Canada are safe. Virtually all factor VIII and most factor IX concentrates used in Canada are made through recombinant technology and are not plasma-derived and thus do not carry the risk of vCJD. Furthermore, plasma-derived products approved for use in Canada by Health Canada are made with plasma from Canada and U.S., where BSE in cattle is extremely rare. Given the extremely low incidence of vCJD in North America and the manufacturing processes designed to remove prions, these plasma derived products are not considered to be at risk.

Since 1998 plasma from UK donors has not been used in the production of any fractionated plasma products. Since 1999 the Canadian Blood Services and Héma-Québec, as well as blood services in the U.S., have not accepted blood and plasma donations from people who have spent specific periods of time in the UK, France and the rest of Europe.

As a result, the Association of Hemophilia Clinic Directors of Canada recommends that NO changes be made to treatment protocols for people with bleeding disorders.

The Canadian Hemophilia Society and the Association of Hemophilia Clinic Directors of Canada will continue to monitor the situation and provide updates.

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For more information

Canadian Hemophilia Society Web site at **www.hemophilia.ca**

World Federation of Hemophilia Web site at **www.wfh.org**

CHANTAL RAYMOND
Canadian Hemophilia Society
1-800-668-2686 | 1-514-848-0503, ext. 226
craymond@hemophilia.ca
www.hemophilia.ca