Event Post mortem finding of asymptomatic variant Creutzfeldt-Jakob Disease abnormal prion protein in a person with haemophilia

Source Haemophilia study carried out by National CJD Surveillance Unit and UK Haemophilia Centres Doctors Organisation

Contact CJD Section, CFI

Background and Interpretation:

A person with haemophilia has been found to have evidence of the prion that causes variant Creutzfeldt-Jakob Disease (vCJD) in his spleen at post mortem. The post mortem was carried out as part of a study jointly co-ordinated by the UK Haemophilia Centre Doctors Organisation (UKHCDO) and the National CJD Surveillance Unit.

This haemophilia patient had been treated in the 1990s with several batches of UK sourced clotting factors, including one batch of factor VIII that was manufactured using plasma from a donor who went on to develop vCJD. The plasma donor developed symptoms of vCJD 6 months after donating the plasma in 1996. The haemophilia patient was in his 70s when he died of a condition unrelated to vCJD, 11 years and one month after receiving the batch of implicated Factor VIII. He had no signs or symptoms of vCJD or other neurological disease when alive.

A final view as to how this haemophilia patient became infected with vCJD has yet to be reached because investigations are continuing to be sure of the aetiology.

This is the first time that a patient with haemophilia, or any patient treated with plasma products, has been found to be infected with vCJD.

All patients with bleeding disorders[1] who have been treated with UK-sourced pooled factor concentrates or antithrombin[2] between 1980 and 2001[3] are classified as at risk of vCJD for public health purposes. Special infection control precautions and other safety measures apply to these patients. This new finding does not change this advice.

This case does not change the public health vCJD 'at risk' status or management of any patients with bleeding disorders.

All haemophilia centre doctors were informed on Monday 16th February 2009. They have been asked to send a letter to all their patients with bleeding disorders as soon as possible.

Implications for HPUs

There will be a press release on this issue on Tuesday 17th February 2009. It is possible that some concerned patients and doctors may contact you for advice. There are no new public health actions to take as a result of this new finding.

Implications for HPA Centres (if appropriate)

Recommendations to HPUs

Please direct enquirers to the HPA website which contains information for patients and healthcare workers. Concerned patients should contact their haemophilia centre. Haemophilia doctors who need further advice should contact the UKHCDO.

References/ Sources of information

Information for doctors and patients will be available from 17/-2/09 at
http://www.hpa.org.uk/vcjdplasmaproducts

[1] Defined here as congenital and acquired haemophilia (Haemophilia A and Haemophilia B), Von Willebrand Disease, other congenital bleeding disorders and congenital antithrombin III deficiency.
[2] ie. clotting factors and antithrombin made from pooled plasma. These include factor VIII, factor IX, factor VII, factor XI, factor XIII and prothrombin complex concentrates as well as antithrombin.
[3] The start date of 1980 is when BSE is thought to have entered the human food chain. The end date of 2001 is the last possible expiry date of any product manufactured by the UK fractionators that was sourced from UK donors until 1998.