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**vCJD ABNORMAL PRION PROTEIN FOUND IN A PATIENT WITH
HAEMOPHILIA AT POST MORTEM**

Evidence of infection with the agent (abnormal prion protein) that causes variant Creutzfeldt-Jakob Disease (vCJD) has been found at post mortem in the spleen of a person with haemophilia.

The patient, who was over 70 years old, died of a condition unrelated to vCJD and had shown no symptoms of vCJD or any other neurological condition prior to his death. The vCJD abnormal prion protein was only identified during post mortem research tests.

The Health Protection Agency is working with the UK Haemophilia Centre Doctors Organisation to ensure all patients with bleeding disorders are made aware of this preliminary information which is being further investigated. This new finding will not change the way patients with haemophilia are cared for or treated.

A final view as to how vCJD abnormal prion protein was transmitted to this haemophilia patient has yet to be reached because investigations are continuing to determine the most likely route of transmission. It is known that the patient had been treated with several batches of UK sourced clotting factors before 1999, which is when measures to improve the safety of blood in relation to vCJD were introduced. The patient's treatment had included one batch of Factor VIII that was manufactured using plasma from a donor who went on to develop symptoms of vCJD six months after donating the plasma in 1996.

This is the first time that vCJD abnormal prion protein has been found in a patient with haemophilia, or any patient treated with plasma products. This new finding, however, does not change the public health vCJD 'at risk' status of patients with bleeding disorders.

Haemophilia patients have previously been informed by their doctors of their possible increased risk of exposure to vCJD via clotting factors. In 2004 all patients with bleeding disorders who had been treated with UK-sourced pooled plasma products between 1980 and 2001 were told that, owing to potential vCJD infectivity from these products they were to be classified as at-risk of vCJD for public health purposes.

Professor Mike Catchpole, Director of the Health Protection Agency's Centre for Infections, said:

"This new finding may indicate that what was until now a theoretical risk may be an actual risk to certain individuals who have received blood plasma products, although the risk could still be quite low. We recognise that this finding will be of concern for

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persons with haemophilia who will be awaiting the completion of the ongoing investigations and their interpretation.

The priority is to ensure that patients are informed of this development and have access to the latest information and specialist advice from their own haemophilia centre doctor as soon as possible.

"This finding does not change our understanding of the risk from vCJD for other people in any specific way. But it does reinforce the importance of the precautionary measures that have been taken over the years.

"Since the risk of vCJD transmission through blood was first considered, a number of precautionary measures have been introduced to minimise the risk from the UK blood supply. UK plasma has not been used for the manufacture of clotting factors since 1999 and synthetic clotting factors are provided for all patients for whom they are suitable."

Ends

Notes for editors

1) The post-mortem tests were carried out as part of a research study jointly coordinated by the UK Haemophilia Centre Doctors Organisation and the National CJD Surveillance Unit. The study was commissioned in 2001 and is ongoing.

2) The likelihood of a person who is infected with the vCJD abnormal prion protein going on to develop symptoms of the disease is uncertain and may depend on individual susceptibility. It is possible that infected individuals may never develop symptoms.

3) Haemophilia is a genetic blood condition in which an essential clotting factor is either partly or completely missing. This causes a person with haemophilia to bleed for longer than normal. Treatment for haemophilia is usually by replacing the missing clotting factor (factor VIII) through regular injections which helps the blood to clot and minimises the likelihood of long term joint damage.

4) In 2004 all patients with bleeding disorders who had been treated with UK-sourced pooled plasma products (e.g. clotting factors for individuals with haemophilia) between 1980 and 2001 were told that, owing to potential vCJD infectivity from these products, they would be classified as at-risk of vCJD for public health purposes.

The start date of 1980 is thought to be the earliest date the agent (abnormal prion protein), that causes BSE in cattle and vCJD in humans, could have entered the food chain. The end date of 2001 is the last possible expiry date of any product manufactured by UK fractionators that had been sourced from UK donors up until 1998.

5) The government introduced a number of measures from 1997 onwards to safeguard blood and plasma supplies.

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- Since 1997 all cases of vCJD that are reported to the National CJD Surveillance Unit and diagnosed as having 'probable' vCJD, result in a search of the UK Blood Services blood donor records. If the patient has donated blood, any unused parts of that blood are immediately removed from stock. The fate of all used components of blood from the donor is traced, and surviving recipients informed of their risk.
- In July 1998, the Department of Health announced that plasma for the manufacture of blood products, such as clotting factors, would be obtained from non-UK sources.
- Since October 1999, white blood cells (which may carry the greatest risk of transmitting vCJD) have been removed from all blood used for transfusion.
- In August 2002 the Department of Health announced that fresh frozen plasma for treating babies and young children born after 1 January 1996 would be obtained from the USA, extended to all children under 16 years of age (Summer 2005).
- In December 2002, the Department of Health completed its purchase of the largest remaining independent US plasma collector, Life Resources Incorporated. This secures long-term supplies of non-UK blood plasma for the benefit of NHS patients.
- Since April 2004, blood donations have not been accepted from people who have themselves received a blood transfusion in the UK since 1980. This has been extended to include apheresis donors and donors who are unsure if they had previously had a blood transfusion (August 2004).
- Since late 2005, blood donations have not been accepted from donors whose blood was transfused to patients who later developed vCJD.
- The UK Blood Services continue to promote the appropriate use of blood and tissues and alternatives throughout the NHS.

6) Specialist advice and care concerning vCJD is available from:

The National CJD Surveillance Unit, based at the Western General Hospital Edinburgh: www.cjd.ed.ac.uk

The NHS National Prion Clinic, based at The Hospital for Neurology and Neurosurgery, Queen Square, London <http://www.nationalprionclinic.org/>

7) For further information about vCJD go to:

www.hpa.org.uk/cjd
<http://www.hpa.org.uk/vcidplasmaproducts>
<http://www.dh.gov.uk/PolicyAndGuidance/HealthAndSocialCareTopics/CJD/fs/en>
<http://www.blood.co.uk/>
<http://www.cjd.ed.ac.uk>
<http://www.nationalprionclinic.org/>

8) For Health Protection Agency media enquiries please contact the Agency's Centre for Infections Press Office on:

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