FOR DECISION

Dawn,
They have suggested that you may wish to MS(PH) write to Lord Archer of Sandwell. Would you like to?
GRO-C

Yes good idea
Agree handling

RESTRICTED- MEDICAL

GRO-C

GRO-C 5/2/09

From: Mark Noterman
Cleared: Liz Woodeson
Date: 3 February 2009
Cc: see list at end.

vCJD PRION PROTEIN FOUND IN HAEMOPHILIAC AT POST MORTEM

Issue

1. To notify you of the first identification of the abnormal protein associated with vCJD infection in the spleen of a haemophiliac, discovered at post mortem. This may represent the first case of transmission via a blood product.

Timing

2. Routine.

Recommendation

3. That you note this finding, and agree the handling proposals.

Background

4. In 2003 the first case of clinical vCJD in a patient who had received whole blood from a donor who later went on to develop vCJD was reported. Since then, a further two cases of vCJD have been identified in patients who received whole blood from two separate donors who also later developed vCJD. A number of actions have been taken to minimise the risks of transmission of vCJD from whole blood and blood products (see **Annex A**). Patients known to have been exposed to blood or blood products from a donor who themselves later went on to develop vCJD, have where possible been notified of their risk.

5. Although no cases of clinical vCJD have been identified in haemophiliacs, they are recognised to be at potential risk because of their regular and frequent requirements for blood products. The blood product clotting factors used to treat haemophilia patients were sourced from the UK until the introduction of US sourced plasma in 1999. Therefore the CJD Incidents Panel (CJDIP, which advises DH and the NHS on CJD risk management) recommended in 2004 that all patients with bleeding disorders who had received UK sourced products between 1980 and 2001 should be notified and managed as a single "umbrella" group potentially at risk of vCJD for public health purposes. This was irrespective of whether they had received clotting factors derived from a donor known to have developed vCJD. The UK Haemophilia Centre Doctors Organisation (UKHCDO) and the Haemophilia Society endorsed this approach.

6. As a follow up to this decision the National CJD Surveillance Unit (NCJDSU) and the UKHCDO are carrying out a study of tissue samples from patients with haemophilia, testing for the abnormal protein associated with vCJD in residual biopsy and autopsy samples.

7. Towards the end of 2008 an autopsy sample from the spleen of a haemophilia patient gave a positive test result. This single positive result was confirmed on repeat testing of that particular sample. A further 23 samples from his spleen, and other tissue samples (brain, appendix, lymph node etc.) from the patient were all negative. The NCJDSU have audited the sampling and testing process, and

consider this single result to be a true positive. The patient was 74 years old when he died, and did not shown any symptoms of vCJD or other neurological disease when alive.

8. The patient's genotype was MV at codon 129. This is a different genotype from all known clinical cases of vCJD (where the genotype is MM), though it is the same as one other case in which the abnormal protein was found at autopsy in the spleen of an elderly asymptomatic female who had received a whole blood transfusion from a donor who later went onto develop clinical vCJD.

9. The haemophilia patient had been treated with a batch of blood product (Factor VIII) prepared from plasma donated by one of the three donors (identified in para 4 above) who later developed vCJD. This plasma was donated in May 1996, and the haemophilia patient died in 2007, 11 years after receiving the Factor VIII.

10. The haemophilia patient had also received blood transfusions from several donors. An initial search of electronic hospital records show that the patient received blood from 14 donors since 1998. The UK Haemophilia Centre Doctors Organisation is leading a case record investigation of the patient's transfusion records and this is expected to reveal further, earlier blood transfusions. The patient is currently not known to have received any whole blood transfusions from any donor who later developed vCJD. The patient is also understood to have several surgical interventions (evidence is being gathered) and if a meat eater will have also have been potentially exposed to the abnormal protein via the dietary route. Once all details are collected analysis will be made of the most likely route of exposure.

Actions

11. Whilst this finding is not unexpected and, subject to the investigation of fuller records, has no immediate implications for the management of haemophilia patients it will be of concern to these patients. There is significant public and Parliamentary interest in blood safety issues, especially in relation to haemophiliacs who have received UK sourced blood products.

12. Pending further advice from the CJD Incidents Panel we have agreed with NHSBT that the National Blood Service (NBS) should continue to accept blood donations from any donors to the haemophilia patient who are still giving blood but not issue them. We have also asked that steps should also be taken to ensure that all identifiable donors to the haemophilia patient are investigated by the National Blood Service and the NCJDSU, to ensure that none of them has been identified as a vCJD clinical case.

13. The Health Protection Agency (HPA) and the UK Haemophilia Centre Doctors Organisation are preparing letters to all Haemophilia Centre Doctors informing them of the case and its implications. The letters will include a core script letter for them to personalise to send to each of the patients "at risk" under their care. The letters to Haemophilia Centre Doctors/haemophilia patients will be sent in March to allow time for more work to be carried out to reduce uncertainty over the most likely route of exposure. This issue of the letters will precede publication of a case report by the NCJDSU.

14. Information on the case was presented to the CJDIP on 22 January, though without the further information awaited on other possible routes of exposure. The panel

formally approved the precautionary actions put in place and proposed by HPA, NHSBT and the UKHCDO, set out in paragraphs 12 and 13 above.

15. Further information about the patient's possible exposure history should be available for the next meeting of the CJD Incidents Panel in May 2009. The Incidents Panel will be asked to consider in the light of this whether:

- any action is needed regarding the 14 or more blood donors to the haemophilia patient;
- there is any change to the risk status of haemophilia patients who received blood product clotting factors sourced a) from the same donor as this patient, and b) from other donors who subsequently developed vCJD;
- there is any change to the risk status for haemophilia patients who received UK sourced blood product clotting factors (unlikely since this does not add new information to the risk assessments which took this possibility into account); and
- there is any change risk status of patients who have received other plasma products such as albumin and immunoglobulin (unlikely since level of risk has not changed).

16. Information on the case will also be presented to the Spongiform Encephalopathy Advisory Committee (SEAC), in confidence, in March and to the Advisory Committee on the Safety of Blood, Tissues and Organs (SaBTO) in April.

Handling, including stakeholder engagement

17. The NCJDSU have agreed not to make their findings public until after the relevant clinicians and patients have been informed.

18. We will prepare a proactive statement and media strategy to accompany the publication of the NCJDSU's report of the case, taking into account any further information available. We do not recommend a Ministerial statement to the house (as with the first possible clinical case of transmission through whole blood, which was a clear new event), because this is not unexpected and all our previous precautionary risk reduction actions have been based on the possibility that it could happen.

Do you want to write to Lord Archer?

19. In view of the ongoing parliamentary and other interest in blood products contaminated in the past, including Lord Archer's independent inquiry into hepatitis C, you may wish to write to Lord Archer, the All Party Parliamentary Group on Haemophilia and the Haemophilia Society immediately in advance of publication of the report of the finding.

Conclusion

20. You are asked:

- to note this case and the proposed action to be taken to inform Haemophilia Centre doctors, and through them their patients, and the issues for further consideration by the CJD Incidents Panel; and
- to agree the handling proposals.

Mark Noterman
Health Protection Division

Cc:
Sarah Kirby
Penelope Irving
Marc McGonagle
Clare Montagu
Marion Dunn
Aimee Gasston
Beatrix Sneller
Sally Davies
David Harper
Liz Woodeson
Ailsa Wight
Peter Bennett
Elaine Gadd
Rowena Jecock
Ben Cole
Sian Jarvis
Kate Pike

Annex A

Current risk reduction measures in place to reduce the risk of vCJD transmission via blood/blood products.

Since the theoretical risk of vCJD transmission through blood was first considered, precautionary measures have been introduced to minimise the risk, including:

Applicable to all blood/blood products

- From December 1997, blood components, plasma products or tissues obtained from any individual who later develops vCJD, have been withdrawn/recalled.

  From October 1999, white blood cells (which may carry a significant risk of transmitting vCJD) have been reduced in all blood used for transfusion, a process known as leucodepletion or leucoreduction.

- Following the report of the first possible case of transmission of vCJD by blood transfusion in December 2003, it was announced in March 2004 that individuals, who had themselves received a transfusion of whole blood components since January 1980, would be excluded from donating blood.

  This exclusion has been extended to include previously transfused platelet donors and donors who are unsure if they have previously had a blood transfusion. This now applies to donors who have been transfused anywhere in the world.

- In July 2004, the exclusion criteria for blood donation were extended to include two new groups, who had received transfusions of whole blood components since 1980:
  - Previously transfused platelet donors,
  - Donors who were unsure if they had previously had a blood transfusion.

- In July 2005, the Department of Health announced further precautionary measures for around 100 patients who donated blood to three people who later developed vCJD. The notified patients have been asked not to donate blood, tissues or organs and to inform health care professionals so extra precautions can be taken when they have surgery or other invasive procedures.

- In November 2005, the Department of Health announced an extension of the notification exercise in July 2005. A further 50 people who had received blood from some of the 100 or so donors notified since July are being traced and notified of their potential exposure to vCJD.

Platelets

To reduce the need to pool donations in producing platelets collection of platelets by apheresis continues to be extended where possible

Plasma

- In July 1998, it was announced that plasma for the manufacture of blood products, such as clotting factors, would be obtained from non-UK sources.

- In August 2002 it was announced that fresh frozen plasma for treating babies and young children born on or after 1 January 1996 would be obtained from the USA.

  Fresh frozen plasma for treating babies and young children born on or after 1 January 1996 is obtained from the USA, and from July 2005 its use was extended to all children up to the age of 16.

  The NHS has been instructed to purchase imported solvent detergent FFP for adult patients with thrombotic thrombocytopenic purpura (TTP), although there is some doubt about the effectiveness of this measure, and further advice will be sought from SaBTO at its next meeting.

Cryoprecipitate

Cryoprecipitate produced from methylene blue treated-plasma imported from the USA is being implemented for all children up to the age of 16.

Additionally, considerable effort is being extended to promote appropriate use of blood throughout the NHS, to target blood use to where it is clinically essential. This work has already achieved notable successes, especially in reducing the use of blood in surgery.