**MEETING OF UK HAEMOPHILLIA DIRECTORS ORGANISATION TO DISCUSS nv CJD AND RISK TO HAEMOPHILLIACS**

**20 NOVEMBER 1997**
**RUSSELL HOTEL, LONDON**

**PRESENT**: Haemophilia Directors
Representatives of DoH and Medicines Control Agency
Representatives of manufacturers

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| Professor John Pattison | - Chairman of SEAC |
| Dr Bob Will | - Director of the CJD Surveillance Centre, Edinburgh |
| Dr Martin Schweiger | - Representing Faculty of Public Health Medicine |
| Dr Angela Robinson | - Director, National Blood Authority |

**Context of Meeting**

1. Guidelines on therapeutic products to treat haemophilia, produced 3 October 1996, that recommend selective use of recombinant Factor 8 have run into difficulties as various purchasers are reluctant to meet the increased costs of treating haemophiliacs (can be 25% to 50% more expensive).

2. The Spongiform Encephalopathy Advisory Committee (SEAC) issued a statement early in November 1997that blood and blood products carry a possible risk of transmitting nvCJD, and recommend that a formal risk assessment is carried out. Even for a rare disease, patients who receive multiple infusions of blood products from large pools (20-65,000 donors) are understandably concerned as are those trying to manage their illness.

   Professor Pattison set out the basis for the advice on blood:

   - The transmissible agent of BSE in cattle is indistinguishable from that of nvCJD (or human BSE)
   - It is not yet possible to predict size or duration of the nvCJD epidemic
   - There is concern that nvCJD could start recycling in the human population as a consequence of blood transfusion since there is no species barrier to cross
   - nvCJD is clinically distinct from classical CJD (cCJD)
   - nvCJD seems to have a greater involvement of the lympho reticular system than cCJD. In scrapie the spleen is affected earlier than the brain, but in the later stage of the disease the brain is more infectious.
   - Emerging evidence suggests that theB lymphocyte may be essential to the pathogenesis of TSEs.

- Thus it would be prudent to minimise any exposure to other people's white cells

- There are no screening questions for donors that are relevant for those asymptomatics who are incubating nvCJD.
- TSEs have been transmitted occasionally by blood products and its components in experimental situations

Iatrogenic CJD has occurred with implants, growth hormone and grafts. Average incubation period is 13 years with a range of 5-25 years

Evidence of cCJD/nvCJD in haemophiliacs is negative at present on the basis of case/control studies, look-back exercises and examination of brains from known haemophiliacs. Of people dying from cCJD, one in six had been a blood donor.

No non-invasive tests suitable for widespread use are available for BSE in cattle, orcCJD/nvCJD in humans or tissues. Diagnosis is based on the clinical picture, histology and capacity of tissues to be infective.

Many factors may influence susceptibility and incubation period.
It is not known if the infective agent acts in a cumulative manner to reach the infection threshold or if an individual dose needs to exceed the individual's infectivity threshold.

White cells can be filtered out but more work needs to be done on how effective this is for B lymphocytes. Filters may damage the lymphocyte in such a way that infective fragments may get through.

An expert group is being convened by the DoH to make a risk assessment (on risk to recipients of blood products) which will report early in 1998. The risk assessment has to be made with many unknowns at each step in the process.

The cost of recombinant Factor 8 is increased by VAT, which is not payable on the highly purified forms already available.

There is considerable anxiety about the issue among haemophiliacs and those caring for them They have already had the HIV disaster.

Issues for the Faculty of Public Health Medicine

- Can we acknowledge that haemophiliacs now face a new hazard, the size of the risk is unquantifiable, but increases with each succeeding infusion of Factor 8?

- Can we support calls to remove VAT from recombinant Factor 8 so that cost is less of a barrier to those wishing to access recombinant Factor 8?

- How do we support those providing services for haemophiliacs in a changing risk environment?

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