

Safety Issues Alerts

Signs of Variant Creutzfeldt-Jakob Disease Found in a UK Patient – NHF Med. Alert #409
<http://www.hemophilia.org/NHFWeb/MainPgs/MainNHF.aspx?menuid=116&contentid=1240>

Medical professionals prospective:

Letter to the Editor Regarding vCJD – BDFW's *The Missing Factor* Vol. 2 Issue 1

Recent news from the United Kingdom regarding the death of an elderly gentleman whose post-mortem examination revealed variant CJD has given rise to renewed concerns and questions about the risk of this disease for patients with bleeding disorders. Given the relatively recent history of viral contamination of blood and blood products, it is understandable that this community would have heightened awareness concern and vigilance regarding any new, potential risks.

In light of this most recent discovery, I would like to present what is know and not known about this case as well as the disease and its transmission. The deceased had received treatment for his hemophilia with a plasma derived factor VIII product that had been obtained from donors prior to 1999 when the vCJD blood safety measures were enacted. A donor to one of these batches developed symptoms of the disease six months after his donation in 1996. This would constitute one possible exposure.

It is also worth remembering that over the span of this man's life in England that he consumed beef raised in the United Kingdom. The World Health Organization's conclusion for the most likely cause of vCJD is exposure to bovine spongiform encephalopathy (BSE) agent and "most plausibly due to dietary contamination by affected bovine central nervous system tissue." His meat consumption during the years prior to the implementation of safeguards would offer another significant risk factor for having contracted the prion. There is no evidence that this patient demonstrated any symptoms of vCJD before his death from other causes. It was only on the required post-mortem that the prion known to cause vCJD was found in his spleen.

It is not clear if this latest case constitutes a change from theoretical risk to an actual risk particularly in light of the fact that safety measures have been in place for nearly a decade. Even when a person is exposed to the prion, it is not well understood who will develop symptoms and who will not. This particular man did not exhibit any symptoms, but there is much that is yet to be learned about the disease through ongoing investigations and surveillance. The discovery of the causative prion for vCJD in this man's spleen reminds us to remain calm in our vigilance, rely upon scientific sources rather than rumor and in the words of my southern friends. "Don't borrow trouble."

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