UK PIN
Advancing Care in Primary Immunodeficiency

UKPIN Statement on vCJD Risk in recipients of Normal Human Immunoglobulin

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Antibody deficient patients will often require endoscopic procedures such as bronchoscopy and GI endoscopy in order to examine for infection and potentially take biopsies looking for inflammatory or neoplastic disease.

Variant CJD (vCJD) was first identified in the UK in 1996, and a total of 178 cases have been reported in the UK to date. Most of these cases have been linked to dietary exposure to bovine spongiform encephalopathy (BSE) in cattle. However three clinical vCJD cases and one case of asymptomatic infection are thought to have occurred as a result of blood-transfusion related person to person spread of infection. A further instance of asymptomatic vCJD infection has been identified in a recipient of UK-sourced clotting factors who had died from other causes [1]. All had received non-leucodepleted red cell transfusions or blood-products manufactured in 1999 or earlier from UK donors who were later diagnosed with clinical vCJD.

Many patients with antibody deficiency will be treated with replacement immunoglobulin. Each batch of immunoglobulin is made from the pooled plasma of not less than 1000 donations. UK plasma has not been used to manufacture immunoglobulin since 1999, in view of the risk of vCJD transmission through blood and blood products [2].

Despite the low risk of transmission of vCJD, which has been further reduced by ceasing to use UK plasma for the manufacture of fractionated plasma products, it has remained standard practice within the UK, on initiating immunoglobulin replacement, to consent patients to the risk of known and unknown infectious agents, often specifically mentioning vCJD.

Guidance with regard to minimising transmission risk of vCJD in healthcare settings is provided by the Advisory Committee of Dangerous Pathogens (ACDP) [4].

No antibody deficient patients exposed to UK sourced immunoglobulin have either developed vCJD or been identified as having evidence of asymptomatic infection to date [1,3]. Moreover, no antibody deficient patients are considered at risk of vCJD as a result of their treatment with UK-sourced immunoglobulin.

Patients who received UK-sourced immunoglobulin should not, as a consequence, be treated any differently from others who are not at risk, with regard to endoscopic or any other procedures. This is a view shared by the United Kingdom Primary Immunodeficiency Network (UKPIN) and the National CJD Research & Surveillance Unit (NCJDRSU), as well as with international experts in immunodeficiency [5].

[1] http://www.cjd.ed.ac.uk/sites/default/files/report25.pdf, accessed 31/10/17

[2] https://www.transfusionguidelines.org/document-library/documents/jpac-position-statement-vcjd-may-2015/download-file/Position%20Statement%20on%20vCJD%20May%202015.pdf, accessed 31/10/17

[3] Helbert MR, Bangs C, Bishop M, Molesworth A, Ironside J. No evidence of asymptomatic variant CJD infection in immunodeficiency patients treated with UK-sourced immunoglobulin. Vox Sang. 2016 Apr;110(3):282-4

[4] https://www.gov.uk/government/publications/guidance-from-the-acdp-tse-risk-management-subgroup-formerly-tse-working-group, accessed 31/10/17

[5] http://www.aaaai.org/ask-the-expert/creutzfeld-jakob, accessed 31/10/17