5.4: Variant Creutzfeldt–Jakob disease (vCJD)

This fatal neurological disease, due to the same agent (abnormal variant of prion protein) as bovine spongiform encephalopathy (BSE) in cattle and caused by eating beef from affected animals, was first identified in the UK in 1996. By the end of 2012 there had been 174 cases in the UK, peaking in 2000. Four cases of transfusion-transmitted vCJD infection have been identified, from three apparently healthy donors who later developed vCJD. All occurred with non-leucodepleted red cells donated before 1999. Three of the four recipients died of vCJD a few years after the implicated transfusion. The fourth recipient died of unrelated causes but had abnormal prion protein in the spleen at post-mortem examination (significance uncertain). There are still many uncertainties around the pathogenesis and epidemiology of vCJD and no practical screening test for blood donors has yet been developed. The vCJD risk-reduction measures introduced in the UK include (see also Chapter 3):

- Importation of plasma for fractionated blood products (1998)
- Leucodepletion of all blood components (1999)
- Importation (and viral inactivation) of fresh frozen plasma for all patients born on or after 1 January 1996 (when dietary transmission of vCJD is assumed to have ceased) (2002)
- Exclusion of blood donors who have received a blood transfusion in the UK since 1980 (2004)

The efficacy and safety of prion filters for blood components has been investigated but their cost-effectiveness is uncertain as the numbers of clinical cases of vCJD have reduced. There is also interest in the cohort of individuals born after measures to eliminate contaminated beef products from the UK diet were instituted in 1996 (‘class of 96’) who are becoming eligible to donate blood as they reach the age of 17.