New variant CJD and the blood supply: Canadian Blood Services reacts

Last September we urged Canadian blood authorities to address the problem of possible transmission of new variant Creutzfeldt–Jakob disease (nvCJD) through the blood supply.[1] We take some small measure of satisfaction in the fact that Canadian Blood Services has finally established an advisory panel to come to terms with this issue.[2] The question is whether any Canadian blood donors who travelled to England or France during the 1980s unknowingly contracted nvCJD through its presumed vector, beef and beef products contaminated with bovine spongiform encephalopathy. The related question of whether nvCJD can be transmitted through the blood supply is still unresolved, although evidence from animal experiments suggests that this possibility cannot be discounted.

Canadian Blood Services estimates that 22% of Canadian donors travelled to the UK during the risk period and 15% to France. Most of these donors give blood frequently. Clearly, deferring their donations would seriously deplete blood supplies and necessitate either a huge increase in donations from other Canadians or the purchase of blood products on the open market, with all of the risks and uncertainties that this would entail. The advisory panel is expected to make its recommendations this month.

New cases of nvCJD continue to surface in the UK. As of Mar. 31, 1999, there were 38 confirmed and 2 probable cases. The median age of onset for the cases reported to the end of 1998 was 28 years; the median age at death was 29 years. (The median age at death for sporadic CJD is 65 years.) The median delay beween onset of symptoms and diagnosis is 15 months. The interval between exposure to the causative agent and confirmation of diagnosis is presumably longer and is the dangerous window for the blood supply. Although the annual number of cases has risen from 3 in 1995 to 16 in 1998, there is no evidence of a real increase in incidence. Some of the observed increase may simply be an artifact of better surveillance.

The diagnosis of nvCJD is extremely difficult to make in the early stages. Dr. R.G. Will of the National Creutzfeldt–Jakob Disease Surveillance Unit in Edinburgh advised neurologists in the UK that the presentation is predominantly psychiatric, involving anxiety, depression, withdrawal and progressive behavioural change: "Nearly all these patients were referred to a psychiatrist early in the clinical course. … After a period of weeks or months [they experienced] a cerebellar syndrome with gait and limb ataxia. Forgetfulness and memory disturbance develop[ed], often late in the clinical course but progress[ed] with the development of severe cognitive impairment and a state of akinetic mutism in the majority of cases."[3]

Close to 600 000 Canadians travel to the UK each year, and hence millions of visits were made during the risk period. Although there have been no confirmed cases of nvCJD in North America, or in fact anywhere outside the UK or France (where a single case has been reported), Canadian physicians should remain alert to this diagnosis.

References

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3. Will RG. Letter to neurologists, Mar. 21, 1996. Available: www.cjd.ed.ac.uk/letter.htm