Blood can transmit Creutzfeldt-Jakob disease

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Creutzfeldt-Jakob disease (CJD) is a rare but fatal disease in humans. For the first time, the presence of infectivity in the blood of patients affected by sporadic and the new variant of CJD was established by scientists from the French National Institute for Agricultural Research (INRA) and the French National Veterinary School (ENVT), in collaboration with European partners. Complementary investigations are underway, but the available results support the contention that CJD might be transmitted by blood transfusion and/or the use of blood derived products. These results are published in the journal *Emerging Infectious Diseases* on 11 December 2013.

Creutzfeldt-Jakob disease (CJD) is a neurodegenerative disorder caused by the accumulation of a misfolded protein, called prion, in the tissues of affected individuals. In humans, there are different forms of the disease. A familial form (genetic origin), a sporadic form (unknown origin) and a form called 'variant', appeared in 1996 as a consequence of dietary exposure to the Bovine Spongiform Encephalopathy agent (BSE or 'mad cow disease').

Several countries (France, Uk, USA, Japan…) have reported that the use of a growth hormone or meninx graft, originating from patients affected or incubating the disease, were responsible for the transmission between individuals of CJD. In the UK, based on epidemiological data, at least four variant CJD contaminations were considered to be the consequence of the transfusion of non leuko-depleted blood units that had been prepared from donorsincubating the disease.

Breaking result: the blood of patients affected by CJD carries infectivity

A team of scientists from INRA and ENV Toulouse (France), in collaboration with the Georg August University (Germany) and the Centro de Investigación en Sanidad Animal (Spain), characterized the presence and distribution of CJD agents (sporadic and variant forms) in the blood. They quantified, by bioassays, the levels of infectivity associated with different blood fractions from CJD affected patients. The blood cells (white and red blood cells) and the plasma from a variant CJD affected patient contained infectivity. Regarding sporadic CJD, infectivity was detected in the plasma of two out of the four investigated cases.

These experiments demonstrate for the first time the presence of infectivity in the blood of CJD affected patients. The results support the contention that CJD might be transmitted by blood transfusion and/or the use of blood derived products. This work also indicates that the distribution of infectivity and the infectious load in the blood of CJD affected patients are similar to the one reported in various animal models of Prion diseases. This confirms the value of animal models to characterize transmission risk associated with prion disease.

Complementary studies involving a greater number of CJD cases are being carried out. Meanwhile, the available results shall allow a refinement of the quantitative risk assessment models related to the CJD blood-borne transmission risk.

More information: Jean Yves Douet, Saima Zafar, Armand Perret-Liaudet, Caroline Lacroux, Séverine Lugan, Naima Aron, Herve Cassard, Claudia Ponto, Fabien Corbière, Juan Maria Torres, Inga Zerr, and Olivier Andreoletti. "Detection of Infectivity in Blood of Persons

with Variant and Sporadic Creutzfeldt-Jakob Disease." *Emerging Infectious Diseases*, online 11 December 2013. DOI: dx.doi.org/10.3201/eid2001.130353