Editorial – The Impact of vCJD on a Safe Supply of Blood and Blood Products

One of the most worrying health problems emerging in the past decades is the epidemic occurrence of new forms of certain neurodegenerative diseases. The prototype of these diseases, a characteristic disorder of sheep and goats called scrapie, had been known for centuries. A similar kind of disease in humans, the Creutzfeldt-Jakob disease (CJD), was first described in 1920. It was found out later that this disease can be transmitted by parenteral application of medicines derived from human substances related to the central nervous system, such as dura mater grafts or hormones from human pituitary glands. Due to these albeit rare transmissions, the typical histologic appearance of involved brain tissue, and the clinical picture, the term transmissible spongiform encephalopathies (TSE) was coined.

About 20 years ago, a new initially mysterious disease of cattle was observed in the UK, which was first called 'mad cow disease'. It was soon found out that it was a new form of TSE, designated bovine spongiform encephalopathy (BSE). It was obviously spread rapidly by feeding material rendered from ruminant carcasses to cattle, resulting in a huge epidemic and tremendous economic losses in the British beef industry. The long-known sheep disease scrapie had never been observed to be transmitted to humans. Thus it was a frightening experience to learn since 1996 that a variant of CJD (vCJD) occurred predominantly in young people, which had obviously to be regarded as a manifestation of BSE in humans.

Many efforts were made within the scientific society to explore and clarify facts concerning origin, course, characteristics and spreading of vCJD. The theory was developed and substantiated that the pathogen behind TSE is a misfolded form of a cellular protein which was named prion. An important task was to develop and optimize methods for detection of prion protein in biologic samples. This is particularly demanding since minuscule amounts of the pathogenic misfolded protein have to be detected in the presence of abundant normal protein. It is also difficult to find appropriate surrogate markers, and there is no detectable immune reaction to TSE. Suitable screening tests, e.g. for blood or organ donors, are not available.

The new disease vCJD brought about a great challenge also for regulatory bodies. This is true not only for the control of the food chain of animals and humans which had to be rigorously cleared from risk materials. Materials derived from cattle are contained in the majority of medicinal products, e.g. as excipients, and precautionary measures were imposed to ensure the safety. After vCJD was first detected, it was immediately clear that a possible secondary infection, i.e. a transmission via human materials used as medicines, had to be considered or even assumed as worst case scenario. Therefore, the task to develop a strategy for a safe blood supply in view of vCJD was on the agenda as a priority issue.

Since the situation with respect to BSE and vCJD epidemiology is very different not only between the continents, but even with Europe, it appears wise that each country should base regulatory decisions on its own risk assessment. Any precautionary measures to ensure blood safety should take into account specific national conditions like the particular BSE epidemiology in cattle, the respective epidemiology in men and the kind and status of national blood product supply.

With the task to develop such a strategy, an expert working group was appointed by the German Federal Minister of Health. The group issued a first report in 2001. The present, substantially revised report provides a summary on prion protein diseases with focus on vCJD and its transmission by blood or blood products. It includes recent developments, e.g. the three cases of vCJD transmission by blood transfusions in the UK, and also modeling studies. Finally, it contains conclusions and recommendations for decision makers with responsibility for blood safety in Germany including the feasibility of certain measures discussed in the last years.

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