PRION 2016 Tokyo Declaration

Prion diseases or transmissible spongiform encephalopathies (TSEs), such as Creutzfeldt-Jakob disease (CJD) in humans, are severe, devastating and intractable diseases. CJD is a rapidly progressive degenerative brain disorder that leads to dementia and other neurological symptoms, resulting in death, usually within months. It is a lethal disease with 100% fatality rate and there is no medicine available to treat the underlying condition. Most CJD cases are sporadic and of unknown origin. There are also hereditary forms such as familial CJD, Gerstmann-Sträussler-Scheinker syndrome and Fatal Familial Insomnia and, rarely, cases have been caused by medical treatments such as human dura mater grafts or human pituitary derived hormones. Prion diseases are due to infectious agents and are found in many animal species, including sheep, cows, deer, and cats, as well as humans.

Prion diseases are believed to start when a normal prion protein converts to a transmissible (infective) abnormal prion protein ("prion"), which then destroys nerve cells. Three Nobel Prizes have been awarded in this narrow field of science (DC Gajdusek 1976, S Prusiner 1997, K Wüthrich 2002), but many aspects of prion diseases, such as how normal host prion proteins are converted to abnormal forms, how they are transmitted and how they make neurons degenerate are far from being fully understood.

Prions can be transmitted through food. In 1996, the world was shocked when the first cases of young people developed variant CJD and this was attributed to having, years earlier, eaten foods contaminated by prion of bovine spongiform encephalopathy (BSE, also known as mad cow disease). Despite a strong mobilization of the scientific community many issues remain uncertain, including the precise origin of BSE prions, the mechanism of infection of young adults through food and whether other animal prion diseases may pose a risk to the human population. This includes atypical BSE and chronic wasting disease in deer, because prions from these animal diseases can be transmitted experimentally to primates or humanized mice, and because, unlike classical BSE, strategies for eliminating these diseases have not yet been established.

In addition, recent studies on proteins that are linked to the main neurodegenerative diseases such as Alzheimer's disease and Parkinson's disease (respectively Aβ protein, Tau protein and α-synuclein protein), have demonstrated these all also share characteristics with prion proteins, notably auto-aggregation, self-propagation and induction of lesions in animals. These findings suggest that these types of neurodegenerative diseases may also be discovered one day as potentially transmissible. They also indicate that research on prion mechanisms can bring unique insight on other devastating diseases, which are common in humans.

We the participants of PRION 2016 including physicians, veterinarians, researchers, scientists, patients, patients' families, industries, and government staff all agree this is the right time to further increase our efforts to overcome prion diseases.

For this purpose, world-wide and intimate cooperation is essential for many reasons including the rarity of the diseases, the evidence of infectivity and the absence of any treatment. The numbers of researchers, physicians, and experts of Prion diseases are limited. In addition, the decline in the number of variant CJD patients and BSE cases in cattle may lead people to misunderstand and wrongly believe that Prion diseases are diseases in the past, whereas prion mechanisms appear to be in fact a critical target in developing new strategies against neurodegenerative diseases.

Here we declare the following statements under the name of PRION 2016 Tokyo Declaration.

Ⅰ. We should aim to increase and improve the understanding of prion diseases in the general population.

Ⅱ. We should facilitate more research to elucidate prion mechanisms, develop new treatments and finally overcome Prion diseases.

Ⅲ. We should promote more international collaboration to pursuit this goal.

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