Prion diseases are rare progressive neurodegenerative disorders that affect both humans and animals. They are distinguished by long incubation periods, characteristic spongiform (Swiss cheese) changes in brain and spinal cord tissues associated with neuronal loss, and a failure to induce inflammatory response.

These diseases are 100% fatal; they appear to arise spontaneously in humans (sporadic) or the causative factor is found in family genetics (inheritable). However, these diseases can also be transmitted to humans eating meat contaminated with BSE or human neuronal material harboring prions, or by contact with brain-derived therapeutic factors like human growth factor extracted for pituitary glands harvested from cadavers.

Surgical instruments used in neurosurgery on an infected patient but are not properly cleaned can be a vehicle for transmitting this disease (prions are not destroyed by conventional sterilization procedures).

Prions are simple proteins that occur in the brain and serve an undetermined function. They cause disease by becoming misfolded, by coming into contact with an external source of misfolded protein, as a result of a spontaneous misfolding event or as a consequence of a hereditary misfolding liability (mutations). The initial misfolded prion catalyzes the transformation of native prions into misfolded forms which cannot be cleared and begin to clump together (aggregate), eventually leading to neuronal death - death by misshapen protein. Misfolded prions are the simplest infectious agents. They differ from a toxin in that they are transmissible. Theoretically,
one misfolded prion can lead to disease and death. Originally named a slow virus infection because of its decades long incubation period. It was assumed that the simplest communicable disease agent was a virus which contained either RNA or DNA as a central dogma for replication. Studies originating in the 60s and culminating in the 80s identified an aberrant protein as the causative and transmissible agent (for which a second Nobel Prize for this disease was awarded).

The human versions of prion disease are Creutzfeldt-Jakob Disease, Variant Creutzfeldt-Jakob Disease, Gerstmann-Straussler-Scheinker Syndrome, Fatal Familial Insomnia, and Kuru.

The zoonotic versions of prion disease are Bovine Spongiform Encephalopathy (BSE), Chronic Wasting Disease (elk), Scrapie (sheep), Transmissible mink encephalopathy, Feline spongiform encephalopathy and Ungulate spongiform encephalopathy.

Although BSE and scrapie have been recognized for centuries, the human disease was only characterized in the 1920s. It was well known that scrapie was a communicable disease, but only among sheep and goats. The study of a very rare neurological condition, kuru, prevalent among the Fore people of Papua New Guinea in the 1950s and 60s, led to the demonstration that prion disease was transmissible in humans. Cannibalistic partaking of brain tissue of deceased tribal members as a ritualistic part of the funeral preparations was identified as the route of transmission (for which the first Nobel Prize was awarded in this field).

Prion disease is not contagious; there is no evidence to suggest it can be spread from person to person by close contact. Once a person has developed prion disease, central nervous system tissues (brain, spinal cord and eye tissue) are thought to be extremely infectious. However, this is only relevant for those handling infected tissue directly, which does not include caregivers looking after a person with the disease. Infectivity in the rest of the body varies in different types of prion disease, but is generally much less than in brain tissue. People with any form of prion disease are requested not to be blood or organ donors, and are requested to inform their doctor and dentist prior to any invasive medical procedures or dentistry.

Given the rarity of these diseases and their exotic mode of transmission, why should prion disease be a concern for us? The possibility of a community acquired plague of prion disease came into clear focus with the jump of the disease from cattle into people during the mad cow disease (BSE) scare in the late 80s and late 90s. Thousands in Britain were exposed to prion contaminated meat from cattle exposed to BSE in their feed. More than 180,000 cattle were infected and millions of cattle were killed during eradication efforts. Over 200 individuals have thus far developed Variant Creutzfeldt-Jakob Disease directly tied to exposure to contaminated meat. We do not know how many individuals exposed to prion-laced beef will eventually develop v-CJD because of the very long incubation period between initial exposure and disease presentation. Everyone who consumed beef while visiting Britain in the 80s and 90s is at some finite risk of developing Variant Creutzfeldt-Jakob Disease in their lifetime. This is a classic case of a zoonotic disease migrating from animals to humans. Could it happen again? Exposure to American bush meat, elk, with Chronic Wasting Disease could lead to an outbreak of a new variant of human prion disease, assuming CWD can make the jump into humans. The meat and meat byproducts industry is now on high alert for any sign of prion disease in herds. The meat industry and regulators are as
vigilant as possible in preventing prion contaminated product from entering the food chain and appearing on the shelves. The economic costs would be devastating for the industry if they get lackadaisical about this issue. Unfortunately, misfolded prion proteins are quite robust. They can persist in the soil for years, which no doubt explains outbreaks of scrapie in sheep feeding in fields where prior scrapie outbreaks have occurred. Sheep shed prions in feces. A large contamination might happen in the future.

Aberrant prions are a perfect doomsday biological warfare agent. The action may be slow to unfold, but the agent is 100% fatal. It is technologically feasible to place genes for pathogenic prion mutants into vector organisms to target humans. Organisms found populating the human gut like, E. coli or some other microbiome bacteria, virus or fungi, could be genetically modified to generate these altered prions and subsequently reintroduced into the human population. Persistent high exposure to misfolded prions in the bowel might considerably shorten the incubation period for the onset of disease. Ultimately lethal doses of these aberrant prion proteins would eventually extinguish human life on earth.

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