

## Prion PrP<sup>h</sup>

The pathogen responsible for the current pandemic is the novel prion PrP<sup>h</sup>.

Prions are proteins, which in their normal form (PrP<sup>c</sup>) are found on the surface of human cells. However, abnormal forms of this protein are able to convert the normal form into their abnormal folded structure. When these cross the blood brain barrier, severe, rapidly progressing neurological disease occurs, with similarities to the prion disease Creutzfeldt-Jacob.

The characteristics of existant prion diseases affecting humans:

- Extremely rare.
- Spread between humans and animals, but not from human to human.
- Spread mainly via ingestion of brain and spinal cord tissue.
- Have long incubation periods before symptoms occur then rapid progression.
- Incurable.
- Caused by a prion resistant to chemical and autoclave destruction except at extreme temperatures.

The novel strain PrP<sup>h</sup> responsible for the pandemic almost certainly originated in the tiny population of the endangered, forest dwelling bovine, Saola. It most likely spread to humans when hunters with an interest in consuming endangered species travelled to Laos. The hunters returned to globally diverse destinations on international flights, suffering from diarrhoea which they believed to be norovirus.

It is now known that the new prion infecting them resembles a novel genetic form first described in 2014, whereby the abnormal prion protein caused watery stools. On the flights the infection spread to at least sixty people via hand to mouth from contaminated surfaces.. Cases are now widely reported across all continents. The features are:

- Now widespread globally, and although cases are falling through implementation of public information and hygiene programs in the developed world, it is increasing exponentially in third world countries where access to clean drinking water is restricted.
- Spreads to humans in two ways. One being hand to mouth transmission via prions found in symptomatic diarrhoea. Due to the largely indestructible nature of the prion, ordinary methods of sewage treatment mean that it is not destroyed. In developed countries it rarely reaches a high enough concentration in drinking water to achieve infection. However in third world countries where drinking water is routinely contaminated with faecal matter, this concentration is frequently reached. Developed world infections have occurred via hand to mouth transfer and via seawater in swimmers exposed to sewage pollution.

- Has a very short incubation period of a few weeks, due to its rapid ability to cross blood brain barrier. This is followed by a quick progression to the symptoms of the bovine to human Creutzfeldt-Jakob disease, i.e. loss of cognition and balance, problems with speech and vision, muscle spasms, progressive dementia and ultimately death.
- Is incurable with no current vaccine. Research is ongoing.
- Is resistant to chemical and normal autoclave destruction. However current research seems to indicate that heat treatment and strong chlorination of water supplies along with microfiltration can prevent transmission, even in areas of high concentration.

### The Populations Affected

The initial individuals infected on flights were from a broad cross section of sexes, ages and races. The fact that the Saola hunters were returning to different continents was unfortunate, as over time this facilitated to a diverse distribution.

Time was lost while scientists sought to explain the novel infection. It was assumed that a prion disease couldn't spread like norovirus or pass on via infected water when in high concentrations. However PrP<sup>Sc</sup>'s protein structure differs, and as well as inducing diarrhoea, it makes the prions less likely to bind to particulate matter in clumps, making it more difficult to remove from drinking water.

The pattern of infection has strongly mimicked that of norovirus. Figures show that around 3% of the population of Europe and North America have died, whilst in parts of Africa, South Asia and South America the figure has been closer to 12%. More children than adults have been affected globally, and this has resulted in parents resorting to measures such as hand binding to stem infection.

Outbreaks in developed countries have tended to centre upon areas of close contact such as schools, hospitals and poor inner city slums. Other clusters of infection in the developed world have come from discharge of untreated sewage into bathing waters. Notable cases in the UK and Albania have happened where bathing water has fallen below EU standards.

PrP<sup>Sc</sup> prion disease adversely culls the poor and the young, and a large percentage of a generation globally is being lost in this unfolding tragedy. Ultimately the loss of this swathe of workers will impact extremely negatively on the older population and western economies reliant on the goods they produce.