Chronic Wasting Disease: Neuro-infectivity as a Result of Oral-fecal Transmission by a Prion

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Chronic wasting disease (CWD) is an infectious disease, caused by a prion, impacting organisms in the Cervidae family (Cervids) (deer elk and moose, and caribou) [1, 2]. Presently, this disease has only been observed in North America and South Korea and occurs by horizontal transmission [2]. Prions are a group of infectious etiological agents categorized as proteinaceous infectious particles which have been shown to lack genetic information (nucleic acids, DNA or RNA) and are biochemically characterized as “misfolded” proteins [3]. Overall, these diseases are in a group called transmissible spongiform encephalopahies (TSE) which result in fatal neurodegenerative disease type events [4, 5].

The first identified prion disease was Scrapie, which was initially classified as a slow virus. It was not until the late 1960’s when this disease (agent) was thought to actually be protein-based [4]. Prion diseases are not restricted to Cervids, they are also found in a wide variety of other organisms, including man [3]. It was not until mRNA of the protein sequence that “true” recognition emerged that these agents were actually proteins (proteinase-resistant prion particles–PrP). This resulted in the concept that “specific” proteins, encoded for the PRPc (cellular prion protein) can misfold becoming an infectious agent (PrPrec) (a β sheet rich conformer) resulting in TSE [4, 5]. How the misfolded protein (prion) then expands the population by “creating” new prions is not completely clear [4]; although, it is recognized infectivity results when a misfolded prion “contacts” a “normal” prion resulting in a conformational change [5]. It is thought PRPc are some form of regulatory neuro-protein and become pathogenic when misfolding occurs (PrPrec). However, a recent caveat suggests not all prions are transmittable indicating there may be different types of prion diseases [4]. This raises the issue of other brain disorders actually being prion-type diseases, but is not recognized as such [5]. It has been shown that susceptibility, even among isoforms of “prions”, can vary [2]. However, the structure of the CWD prion may be the primary reason why this “disease” has not been reported in man; although, consumption of contaminated meat commonly occurs [6]. Amino acid differences may act as a barrier in transferring CWD (PrPcwD) to other species, most notably man and cattle [7].

The origin of CWD is not known but two theories exist regarding its occurrence [2, 8]. Suggestions on the origin of CWD have focused on it jumping species from Scrapie and the second as a spontaneous event. TSE’s in humans are known to spontaneously occur at a rate of about one in a million. CWD was first identified in the late 1960’s in captive deer populations and later, around 1978, in free-ranging animals [2]. This first occurrence was in southern Wyoming/Colorado area. Another concept on the emergence of CWD is that it does periodically spontaneously occur but has been kept in check by predators; thus not observed by those working with wildlife until recently. The loss of predators over the last 100 years may have resulted in the exaggerated occurrence and distribution of this disease. Thus, a spontaneous change of PRPc to PrPcwD may be a “periodic”, although an unusual occurrence, but due to an existence of predators may not be seen nor easily spread into a population. Thus, a spontaneous event in an individual maybe controllable when strong predator pressure exists preventing effective spread into the population. Here predator control may be effective when a spontaneous event occurs, but not for an infected population or after an endemic epiosde occurs. However hampering this idea, investigations [9] have indicated even with high predation there is little impact on spread of this disease in the susceptible population. This could be due to, in part, prions surviving transfer through the digestive tract with predators acting as a mechanism of translocation [10]. This may be occurring even though infected animals appear to have a higher predation rate [11]. With a long survival time in soil and...
even translocation of prions within plants, there may be a positive pressure to maintain this disease in a geographic area [12].

Studies [2, 13] have suggested CWD can be orally-fecal transmitted. There are other mechanisms of potential transfer occurring for wildlife (e.g. saliva, carcass) [13, 14]. Prions are highly resistant to degradation and have been shown to exist in soil for years [1]. Recent studies have reported these agents can bind and be taken up by plants [12]. Further, infectivity can be spread when animals consume contaminated plant material. It appears that in endemic areas urine and feces from infected wildlife contain a high enough concentration of PrP to result in plant and soil contamination [1, 12]. Since soil may serve as a natural reservoir for prions, and plants may translocate these agents, various abiotic and biotic “sources” may be acting as a long-term component preventing elimination of the disease [2, 12, 15]. This raises the question as to what risk exists for man?

Case reports have indicated CWD has been transmitted to man, but these suggestions have not stood up to rigorous investigation [16]. An epidemiological study [17] has indicated that even in an area (Colorado) where CWD has existed for years there does not appear to be an increase in “prion-related” diseases in man. However, CWD remains a concern for those in wildlife management [18]. Reports [2] have shown CWD (prions) exist in muscle of infected organisms. This suggests risk is low regarding transmission and possibly due to genetic differences may actually not be transmissible to man (cross-species transmission) [2, 6, 7]. Protection of man from CWD may be due to requirements for transmission occurring only when certain genetic polymorphisms exist allowing change of the PRPC to the infectious form [6, 7, 19, 20]. Based on amino acid evidence (structural determinants) of CWD and human PrP it appears there is a substantial barrier for disease transmission of this agent to humans [6, 7, 19, 20]. This appears to be the reason why Scrapie is not transmitted to Cervids even though sheep exist in the same geographic location [19]. These “genetic” differences appear to be effective as a species barrier for preventing transmission. Thus, there does not appear to be an actual risk to man of CWD as a result of consuming “contaminated” food products from Cervids.
References