Review

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Amyloids, prions and the inherent infectious nature of misfolded protein aggregates

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Abstract

Misfolded aggregates present in amyloid fibrils are associated with various diseases known as "protein misfolding" disorders. Among them, prion diseases are unique in that the pathology can be transmitted by an infectious process involving an unprecedented agent known as a "prion". Prions are infectious proteins that can transmit biological information by propagating protein misfolding and aggregation. The molecular mechanism of prion conversion has a striking resemblance to the process of amyloid formation, suggesting that misfolded aggregates have an inherent ability to be transmissible. Intriguing recent data suggest that other protein misfolding disorders might also be transmitted by a prion-like infectious process.