Prion Proteins and Sleep Disturbances

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Abstract

Prions normally exist as cellular membrane proteins. In humans, 209 amino acids with one disulfide bond form a primarily alpha-helical prion protein structure with a molecular mass of 35 to 36 kDa. The specific role and function of the prion protein elude research efforts and remains a controversial topic. Misfolding of the native prion protein leads to a protein structure with increased proportion of alpha-helices to beta-sheets. Advancing our understanding of the role of the prion protein as it relates to sleep and sleep disturbances presents an appealing avenue into diagnosing and more effectively treating a devastating and debilitating disease. New research into multiple system atrophy further validates evidence of a direct association between the prion protein and sleep. This reinforces previous observations regarding changes in sleep patterns noted with patients affected by Creutzfeldt-Jakob Disease and Fatal Familial Insomnia. From these earlier studies, a more focused approach to identifying and defining the role of the prion protein appears possible. A clearer understanding of the functional prion protein in its native role within the cell membrane allows identification of the potential signaling pathways and the aberration that likely occurs that leads to misfolding at the thermodynamic level. This discovery holds the greater, global potential of elucidating the mystery of proteopathies.

With difficulty in diagnosing a patient affected by prion disease, an association with sleep and prion appears likely to provide a more effective means for assessment and planning. Currently, definitive diagnosis requires pathologic evaluation of brain tissue and/or genetic testing [3,11]. By identifying definitive abnormalities assessed through current diagnostic techniques and criteria, such as multi-channel polysomnography, accurate diagnosis of prion infection with high sensitivity and specificity appears feasible [11]. This association promises a significant advancement in diagnosis and development of more effective treatments.

Further, identification of a clear association with physiologic mechanisms such as sleep provides a means for focusing research efforts to more effectively identify the role of the prion protein. A more lucid identification of the potential signaling pathway(s) involving the prion promotes development of greater insight into cellular and protein signaling in general. Misfolded proteins are implicated in various diseases to include, but not limited to Alzheimer’s, Parkinson’s, type 2 diabetes and amyloidosis grouped under “proteopathies” [12]. Misfolding of the protein appears likely due to abnormal signaling from protein-protein interaction; however, the specific mechanism remains elusive and currently exists only as a theory.

Advancing our understanding of the role of the prion protein as it relates to sleep and sleep disturbances presents an appealing avenue into diagnosing and more effectively treating a devastating and debilitating disease. New research into MSA further validates evidence of a direct association between the prion protein and sleep. This reinforces previous observations regarding changes in sleep patterns noted with patients affected by CJD and FFI. From these earlier studies, a more focused approach to identifying and defining the role of the prion protein appears possible. A clearer understanding of the functional prion protein in its native role within the cell membrane allows identification of the potential signaling pathways and the
aberration that likely occurs that leads to misfolding at the thermodynamic level. This discovery holds the greater, global potential of elucidating the mystery of proteopathies.

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