

Protein misfolding and disease: The case of prion disorders

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Recent findings strongly support the hypothesis that diverse human disorders, including the most common neurodegenerative diseases, arise from misfolding and aggregation of an underlying protein. Despite the good evidence for the involvement of protein misfolding in disease pathogenesis, the mechanism by which protein conformational changes participate in the disease is still unclear. Among the best-studied diseases of this group are the transmissible spongiform encephalopathies or prion-related disorders, in which misfolding of the normal prion protein plays a key role in the disease. In this article we review recent data on the link between prion protein misfolding and the pathogenesis of spongiform encephalopathies.