Prion propagation in vitro: are we there yet?

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Abstract:

Prion diseases are caused by proteinaceous pathogens termed prions. Although the details of the mechanism of prion propagation are not fully understood, conformational conversion of cellular prion protein (PrPC) to misfolded, disease-associated scrapie prion protein (PrPSc) is considered the essential biochemical event for prion replication. Currently, studying prion replication in vitro is difficult due to the lack of a system which fully recapitulates the in vivo phenomenon. Over the last 15 years, a number of in vitro systems supporting PrPC conversion, PrPSc amplification, or amyloid fibril formation have been established. In this review, we describe the evolving methodology of in vitro prion propagation assays and discuss their ability in reflecting prion propagation in vivo.

Key Word:
prion disease, prion, cellular prion protein, disease-associated scrapie prion protein, in vitro conversion, in vitro prion amplification, prion infectivity

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