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Prions: A Mystery Unravelling?

Abstract. Prions result in fatal degeneration of the central nervous system (CNS) in the form of diseases known as *transmissible spongiform encephalopathies* (TSEs). The discovery in 1996 of a new variant of Creutzfeldt-Jakob disease (a human TSE) and experimental confirmation that it is caused by the prion strain responsible for bovine spongiform encephalopathy (BSE) has greatly spurred research in this field. The mechanism underlying prion propagation is now reasonably clear. Prions multiply, in fact, by stimulating their hosts to produce proteins that are initially normal, but acquire an abnormal, prion-like conformation during the coiling stage. A fuller understanding of this mechanism could lead to the employment of molecules capable of making prion proteins revert to the normal conformation in the treatment of both TSEs and other serious CNS disorders.