

## References

- Aguzzi, A., & Calella, A. M. (2009). Prions: Protein aggregation and infectious diseases. *Physiological Reviews*, *89*(4), 1105–1152.
- Alper, T., Cramp, W. A., Haig, D. A., & Clarke, M. C. (1967). Does the agent of scrapie replicate without nucleic acid? *Nature*, *214*(5090), 764–766.
- Antonyuk, S. V., Trevitt, C. R., Strange, R. W., Jackson, G. S., Sangar, D., Batchelor, M., et al. (2009). Crystal structure of human prion protein bound to a therapeutic antibody. *Proceedings of the National Academy of Sciences of the United States of America*, *106*(8), 2554–2558.
- Bessen, R. A., & Marsh, R. F. (1994). Distinct PrP properties suggest the molecular basis of strain variation in transmissible mink encephalopathy. *Journal of Virology*, *68*(12), 7859–7868.
- Brandner, S., Whitfield, J., Boone, K., Puwa, A., O'Malley, C., Linehan, J. M., et al. (2008). Central and peripheral pathology of kuru: Pathological analysis of a recent case and comparison with other forms of human prion disease. *Philosophical Transactions of the Royal Society of London Series B, Biological Sciences*, *363*(1510), 3755–3763.
- Bremer, J., Baumann, F., Tiberi, C., Wessig, C., Fischer, H., Schwarz, P., et al. (2010). Axonal prion protein is required for peripheral myelin maintenance. *Nature Neuroscience*, *13*(3), 310–318.
- Castilla, J., Morales, R., Saa, P., Barria, M., Gambetti, P., & Soto, C. (2008). Cell-free propagation of prion strains. *The EMBO Journal*, *27*(19), 2557–2566.
- Caughey, B. (2003). Prion protein conversions: Insight into mechanisms, TSE transmission barriers and strains. *British Medical Bulletin*, *66*, 109–120.

- Colby, D. W., Giles, K., Legname, G., Wille, H., Baskakov, I. V., DeArmond, S. J., et al. (2009). Design and construction of diverse mammalian prion strains. *Proceedings of the National Academy of Sciences of the United States of America*, *106*(48), 20417–20422.
- Collinge, J. (1999). Variant Creutzfeldt-Jakob disease. *Lancet*, *354*(9175), 317–323.
- Collinge, J. (2001). Prion diseases of humans and animals: Their causes and molecular basis. *Annual Review of Neuroscience*, *24*, 519–550.
- Collinge, J. (2005). Molecular neurology of prion disease. *Journal of Neurology Neurosurgery and Psychiatry*, *76*(7), 906–919.
- Collinge, J., & Clarke, A. (2007). A general model of prion strains and their pathogenicity. *Science*, *318*(5852), 930–936.
- Collinge, J., Sidle, K. C., Meads, J., Ironside, J., & Hill, A. F. (1996). Molecular analysis of prion strain variation and the etiology of ‘new variant’ CJD. *Nature*, *383*(6602), 685–690.
- Collinge, J., Whitfield, J., McKintosh, E., Beck, J., Mead, S., Thomas, D. J., et al. (2006). Kuru in the 21st century—an acquired human prion disease with very long incubation periods. *Lancet*, *367*(9528), 2068–2074.
- Deleault, N. R., Harris, B. T., Rees, J. R., & Supattapone, S. (2007). Formation of native prions from minimal components *in vitro*. *Proceedings of the National Academy of Sciences of the United States of America*, *104*(23), 9741–9746.
- Griffith, J. S. (1967). Self-replication and scrapie. *Nature*, *215*(5105), 1043–1044.
- Kim, J. I., Cali, I., Surewicz, K., Kong, Q., Raymond, G. J., Atarashi, R., et al. (2010). Mammalian prions generated from bacterially expressed prion protein in the absence of any mammalian cofactors. *The Journal of Biological Chemistry*, *285*(19), 14083–14087.

- Mallucci, G., & Collinge, J. (2005). Rational targeting for prion therapeutics. *Nature Reviews Neuroscience*, 6(1), 23–34.
- Mallucci, G., Dickinson, A., Linehan, J., Klohn, P., Brandner, S., & Collinge, J. (2003). Depleting neuronal PrP in prion infection prevents disease and reverses spongiosis. *Science*, 302(5646), 871–874.
- Mead, S., Poulter, M., Uphill, J., Beck, J., Whitfield, J., Webb, T. E., et al. (2009a). Genetic risk factors for variant Creutzfeldt-Jakob disease: A genome-wide association study. *Lancet Neurology*, 8(1), 57–66.
- Mead, S., Stumpf, M. P., Whitfield, J., Beck, J., Poulter, M., Campbell, T., et al. (2003). Balancing selection at the prion protein gene consistent with prehistoric kuru-like epidemics. *Science*, 300(5619), 640–643.
- Mead, S., Whitfield, J., Poulter, M., Shah, P., Uphill, J., Campbell, T., et al. (2009b). A novel protective prion protein variant that colocalizes with kuru exposure. *The New England Journal of Medicine*, 361(21), 2056–2065.
- Nicoll, A. J., & Collinge, J. (2009). Preventing prion pathogenicity by targeting the cellular prion protein. *Infectious Disorders Drug Targets*, 9(1), 48–57.
- Parchi, P., Castellani, R., Capellari, S., Ghetti, B., Young, K., Chen, S. G., et al. (1996). Molecular basis of phenotypic variability in sporadic Creutzfeldt-Jakob disease. *Annals of Neurology*, 39(6), 767–778.
- Prusiner, S. B. (1982). Novel proteinaceous infectious particles cause scrapie. *Science*, 216(4542), 136–144.
- Prusiner, S. B. (1998). Prions. *Proceedings of the National Academy of Sciences of the United States of America*, 95(23), 13363–13383.

- Riesner, D. (2003). Biochemistry and structure of PrP(C) and PrP(Sc). *British Medical Bulletin*, 66, 21–33.
- Safar, J., Wille, H., Itri, V., Groth, D., Serban, H., Torchia, M., et al. (1998). Eight prion strains have PrP<sup>Sc</sup> molecules with different conformations. *Nature Medicine*, 4(10), 1157–1165.
- Safar, J. G., Kellings, K., Serban, A., Groth, D., Cleaver, J. E., Prusiner, S. B., et al. (2005). Search for a prion-specific nucleic acid. *Journal of Virology*, 79(16), 10796–10806.
- Sigurdson, C. J. (2008). A prion disease of cervids: Chronic wasting disease. *Veterinary Research*, 39(4), 41.
- Silveira, J. R., Raymond, G. J., Hughson, A. G., Race, R. E., Sim, V. L., Hayes, S. F., et al. (2005). The most infectious prion protein particles. *Nature*, 437(7056), 257–261.
- Telling, G. C., Parchi, P., DeArmond, S. J., Cortelli, P., Montagna, P., Gabizon, R., et al. (1996). Evidence for the conformation of the pathologic isoform of the prion protein enciphering and propagating prion diversity. *Science*, 274(5295), 2079–2082.
- Wadsworth, J. D., & Collinge, J. (2007). Update on human prion disease. *Biochimica et Biophysica Acta*, 1772(6), 598–609.
- Wadsworth, J. D., Joiner, S., Linehan, J. M., Asante, E. A., Brandner, S., & Collinge, J. (2008). Review. The origin of the prion agent of kuru: Molecular and biological strain typing. *Philosophical Transactions of the Royal Society of London Series B, Biological Sciences*, 363(1510), 3747–3753.
- Watts, J. C., Drisaldi, B., Ng, V., Yang, J., Strome, B., Horne, P., et al. (2007). The CNS glycoprotein Shadoo has PrP(C)-like protective properties and displays reduced levels in prion infections. *The EMBO Journal*, 26(17), 4038–4050.

Weissmann, C. (1991). A “unified theory” of prion propagation. *Nature*, 352(6337), 679–683.

Weissmann, C., & Flechsig, E. (2003). PrP knock-out and PrP transgenic mice in prion research.

*British Medical Bulletin*, 66, 43–60.

Wickner, R. B., Edskes, H. K., Shewmaker, F., & Nakayashiki, T. (2007). Prions of fungi:

Inherited structures and biological roles. *Nature Reviews Microbiology*, 5(8), 611–618.