

Product Information

Anti-Prion Protein antibody, Mouse monoclonal
clone 8H4, purified from hybridoma cell culture

Catalog Number **P0110**

Product Description

Anti-Prion Protein antibody, Mouse monoclonal (mouse IgG2b isotype) is derived from the hybridoma 8H4 produced by the fusion of mouse myeloma cells (SP2/0 cells) and splenocytes from Prnp^{-/-} mice immunized with recombinant murine PrP^C (Gene ID: 19122).¹ The isotype is determined using a double diffusion immunoassay using Mouse Monoclonal Antibody Isotyping Reagents, Catalog Number ISO2.

Anti-Prion Protein antibody, Mouse monoclonal recognizes human,^{1,3} monkey,¹ cow,¹ sheep,¹ squirrel,¹ mouse,^{1,2} and rat prion (25-35 kDa). The antibody may be used in various immunochemical techniques including: ELISA,¹ immunoblotting,¹ flow cytometry,¹ immunocytochemistry,^{1,3} immunohistochemistry,^{1,2} immunoprecipitation,³ and immunoelectron microscopy.¹ The antibody epitope resides within amino acids 145-180 of human prion.^{1,3}

Prion-related diseases are fatal neurodegenerative disorders also known as transmissible spongiform encephalopathies (TSEs). Such TSEs include Creutzfeldt-Jacob disease (CJD), Gerstmann-Strausler-Scheinker syndrome (GSS) and fatal familial insomnia (FFI) in humans, bovine spongiform encephalopathy (BSE) and scrapie in sheep, and chronic wasting disease in elk.⁴ Histological characteristics of TSEs include spongiform change, astrocytosis, neuronal loss and progressive accumulation of amyloid plaques containing protease-resistant prion protein. The modified state (known as PrP^{Sc} for scrapie-associated prion protein) is the infectious agent and mutated PrP genes are responsible for the hereditary aspect of TSEs.⁵ The root cause of TSEs was thought to be nucleic acids in the form of viral DNA or RNA. However, after exhaustive research into the nature of scrapie infectivity, Prusiner and his colleagues presented the controversial hypotheses that the disease was spread by a "proteinaceous infectious particle" or prion.⁶ The prion protein is a natural protein synthesized within the

secretory pathway and transported to the surface of the cell where it is tethered to the cell membrane by a glycosylphosphatidylinositol (GPI) anchor.^{7,8} PrP is constitutively expressed in brain and other tissues of healthy humans and animals and is present in high levels at the synapse.⁹ The activity of PrP is not well understood; it may be involved in copper utilization,¹⁰ serving to buffer copper at the synaptic cleft or to mediate re-uptake of copper into the presynapse. Alternatively, bound copper may influence PrP binding characteristics. The PrP-copper complex may be crucial for synaptic homeostasis as a result of its antioxidant activity.⁹ Aggregates of prion protein are often, but not always, found in brains of individuals with a prion disease. Prion plaques are of three types: unicentric (single, compact core), multicentric (two or more cores and definite border), and diffuse plaques without a definite central core.¹¹ Disease-associated prion protein specifically inhibits the proteolytic β -subunits of the 26S proteasome. This may clarify the mechanism of cell death by the prion protein.¹²

Reagent

Supplied as a solution in 0.01 M phosphate buffered saline, pH 7.4, containing 15 mM sodium azide as a preservative.

Antibody concentration: ~1.5 mg/mL

Precautions and Disclaimer

This product is for R&D use only, not for drug, household, or other uses. Please consult the Material Safety Data Sheet for information regarding hazards and safe handling practices.

Storage/Stability

For continuous use, store at 2-8 °C for up to one month. For extended storage, freeze in working aliquots. Repeated freezing and thawing, or storage in "frost-free" freezers, is not recommended. If slight turbidity occurs upon prolonged storage, clarify the solution by centrifugation before use. Working dilution samples should be discarded if not used within 12 hours.

Product Profile

Immunoblotting: a working concentration of 2-4 µg/mL is recommended using mouse brain extract.

Note: In order to obtain the best results using various techniques and preparations, we recommend determining optimal working dilutions by titration.

References

1. Zanusso, G., et al., *Proc. Natl. Acad. Sci. USA*, **95**, 8812-8816 (1998).
2. Liu, W.G., et al., *J. His. Cyto.*, **51**, 1065-1071 (2003).
3. Gu, Y., et al., *Mol. Cell. Biol.*, **26**, 2697-2715 (2006).
4. Ironside, J.W., and Bell, J.E., In: *Prion Diseases*, Eds.: Collinge, J., and Palmer, M.S., p. 57-88, Oxford University Press (1997).
5. Prusiner, S.B., et al., *Science*, **252**, 1515-1522 (1991).
6. Prusiner, S.B., et al., *Science*, **216**, 136-144 (1982).
7. Stahl, N., et al., *Cell*, **51**, 229-240 (1987).
8. Caughey, B., et al., *J. Virol.*, **63**, 175-181 (1989).
9. Brown, D.R., *Trends Neurosci.*, **24**, 85-90 (2001).
10. Kretzschmar, H A., et al., *Arch. Virol. Suppl.*, **16**, 239-249 (2000).
11. Rezaie, P., and Lantos, P.L., *Brain Res. Rev.*, **35**, 55-72 (2001).
12. Kristiansen, M., et al., *Mol. Cell.*, **26**, 175-188 (2007).

DS,PHC 01/16-1