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Product Information

MONOCLONAL ANTI-PRION PROTEIN

Clone 3F4

Purified Mouse Immunoglobulin

Product Number **P 1115**

Product Description

Monoclonal Anti-Prion Protein (PrP, mouse IgG2A isotype) is derived from the hybridoma produced by the fusion of mouse myeloma cells and splenocytes from a BALB/c mouse immunized with PrPs derived from strain 263K, a hamster-adapted scrapie strain, as the immunogen. The immunoglobulin is isolated from mouse ascites fluid by Protein A chromatography.

Monoclonal Anti-Prion Protein recognizes amino acid residues 109-112 of PrP from human, hamster and feline by immunoblotting, ELISA, immunoprecipitation and immunohistochemistry on fixed sections. This antibody does not react with PrP from any other mammalian species. Monoclonal Anti-Prion Protein is reactive with both native and denatured forms of PrP. This antibody recognizes both the denatured protease sensitive (PrP^C) and denatured protease insensitive (PrP^{Sc}) forms of PrP.

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.¹ Histological characteristics of TSEs include spongiform change, astrocytosis, neuronal loss and progressive accumulation of amyloid plaques containing protease-resistant prion protein.

The endogenous prion protein (known as PrP^C for cellular prion protein), in a 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.² The root cause of TSEs was thought to be nucleic acid 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.³

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.^{4,5} PrP is constitutively expressed in brain and other tissues of healthy humans and animals and is present in high levels at the synapse.⁶ The activity of PrP is not well understood; it may be involved in copper utilization,⁷ 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 anti-oxidant activity.⁶

Aggregates of prion protein are often, but not always, found in the brains of individuals with a prion disease. Prion plaques occur in three types: unicentric (single, compact core), multicentric (two or more cores and definite border), and diffuse plaques without a definite central core.⁸

A tremendous amount of research remains to be done before the prion diseases are understood. An understanding of the mechanism underlying these diseases will not only lead to potential new therapies, but will also answer more fundamental questions concerning protein conformation and architecture in cell biology.

Reagent

Monoclonal Anti-Prion Protein is supplied as 100 μ l of purified immunoglobulin at 1 mg/ml in phosphate buffered saline with 0.03 % thimerosal as preservative.

Precautions and Disclaimer

Due to the thimerosal content, a material safety data sheet (MSDS) for this product has been sent to the attention of the safety officer of your institution. Consult the MSDS for information regarding hazardous and safe handling.

Storage/Stability

Store at -20 °C.

For extended storage, freeze in working aliquots. Repeated freezing and thawing is not recommended. 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

The recommended working dilutions are 1:10⁵ to 1:10⁶ for ELISA; 1:10⁴ to 1:10⁵ for immunoblotting; 1:10 to 1:100 for immunoprecipitation, and 1:100 to 1:1000 for immunohistochemistry.

For the purpose of immunohistochemistry, the epitope must be re-exposed in fixed tissues by pretreatment of tissue with either hydrolytic autoclaving or with 70 % formic acid for 10 minutes at room temperature.

Note: In order to obtain best results and assay sensitivities of different techniques and preparations, we recommend determining optimal working dilutions by titration test.

References

1. Ironside, J. W. and Bell, J. E., Pathology of Prion Diseases. In Prion Diseases, Collinge, J. and Palmer, M. S. (Eds.) pp. 57-88 (Oxford University Press, 1997).
2. Prusiner, S. B., et al., Science, **252**, 1515-1522 (1991).
3. Prusiner, S. B., et al., Science, **216**, 136-144 (1982).
4. Stahl, N., et al., Cell, **51**, 229-240 (1987).
5. Caughey, B., et al., J. Virol., **63**, 175-181 (1989).
6. Brown, D. R., Trends Neurosci., **24**, 85-90 (2001).
7. Kretzschmar, H. A. et al., Arch. Virol. Suppl., **16**, 239-249 (2000).
8. Rezaie, P. and Lantos, P. L., Brain Res. Rev., **35**, 55-72 (2001).

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