

PRNP Recombinant Rabbit mAb

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Catalog # AP94218

Product Information

Application	WB, IHC-P, IHC-F, IF
Reactivity	Rat, Human, Mouse
Host	Rabbit
Clonality	Recombinant
Physical State	Liquid
Immunogen	KLH conjugated synthetic peptide derived from PRNP
Epitope Specificity	161-253 / 253
Isotype	IgG
Purity	affinity purified by Protein A
Buffer	0.01M TBS (pH7.4) with 1% BSA, 0.02% Proclin300 and 50% Glycerol.
SUBCELLULAR LOCATION	Cell membrane. Golgi apparatus and Cytoplasm. Nucleus. Accumulates outside the secretory route in the cytoplasm, from where it relocates to the nucleus.
DISEASE	Note=PrP is found in high quantity in the brain of humans and animals infected with neurodegenerative diseases known as transmissible spongiform encephalopathies or prion diseases, like: Creutzfeldt-Jakob disease (CJD), fatal familial insomnia (FFI), Gerstmann-Straussler disease (GSD), Huntington disease-like type 1 (HDL1) and kuru in humans; scrapie in sheep and goat; bovine spongiform encephalopathy (BSE) in cattle; transmissible mink encephalopathy (TME); chronic wasting disease (CWD) of mule deer and elk; feline spongiform encephalopathy (FSE) in cats and exotic ungulate encephalopathy (EUE) in nyala and greater kudu. The prion diseases illustrate three manifestations of CNS degeneration: (1) infectious (2) sporadic and (3) dominantly inherited forms. TME, CWD, BSE, FSE, EUE are all thought to occur after consumption of prion-infected foodstuffs. Defects in PRNP are the cause of Creutzfeldt-Jakob disease (CJD) [MIM:123400]. CJD occurs primarily as a sporadic disorder (1 per million), while 10-15% are familial. Accidental transmission of CJD to humans appears to be iatrogenic (contaminated human growth hormone (HGH), corneal transplantation, electroencephalographic electrode implantation, etc.). Epidemiologic studies have failed to implicate the ingestion of infected animal meat in the pathogenesis of CJD in human. The triad of microscopic features that characterize the prion diseases consists of (1) spongiform degeneration of neurons, (2) severe astrocytic gliosis that often appears to be out of proportion to the degree of nerve cell loss, and (3) amyloid plaque formation. CJD is characterized by progressive dementia and myoclonic seizures, affecting adults in mid-life. Some patients present sleep disorders, abnormalities of high cortical function, cerebellar and corticospinal disturbances. The disease ends in death after a 3-12 months illness. Defects in PRNP are the cause of fatal familial insomnia (FFI) [MIM:600072]. FFI is an autosomal dominant disorder and is characterized by neuronal degeneration limited to selected thalamic nuclei and progressive insomnia. Defects in PRNP are the cause of Gerstmann-Straussler disease (GSD) [MIM:137440]. GSD is a

heterogeneous disorder and was defined as a spinocerebellar ataxia with dementia and plaquelike deposits. GSD incidence is less than 2 per 100 million live births. Defects in PRNP are the cause of Huntington disease-like type 1 (HDL1) [MIM:603218]. HDL1 is an autosomal dominant, early onset neurodegenerative disorder with prominent psychiatric features. Defects in PRNP are the cause of kuru (KURU) [MIM:245300]. Kuru is transmitted during ritualistic cannibalism, among natives of the New Guinea highlands. Patients exhibit various movement disorders like cerebellar abnormalities, rigidity of the limbs, and clonus. Emotional lability is present, and dementia is conspicuously absent. Death usually occurs from 3 to 12 month after onset. Defects in PRNP are the cause of spongiform encephalopathy with neuropsychiatric features (SENF) [MIM:606688]; an autosomal dominant presenile dementia with a rapidly progressive and protracted clinical course. The dementia was characterized clinically by frontotemporal features, including early personality changes. Some patients had memory loss, several showed aggressiveness, hyperorality and verbal stereotypy, others had parkinsonian symptoms. Prion diseases, or transmissible spongiform encephalopathies (TSEs), are manifested as genetic, infectious or sporadic, lethal neurodegenerative disorders involving alterations of the prion protein (PrP). Characteristic of prion diseases, cellular PrP (PrP_c) is converted to the disease form, PrP_{Sc}, through alterations in the protein folding conformations. PrP_c is constitutively expressed in normal adult brain and is sensitive to proteinase K digestion, while the altered PrP_{Sc} conformation is resistant to proteases, resulting in a distinct molecular mass after PK treatment. Consistent with the transient infection process of prion diseases, incubation of PrP_c with PrP_{Sc} both in vitro and in vivo produces PrP_c that is resistant to protease degradation. Infectious PrP_{Sc} is found at high levels in the brains of animals affected by TSEs, including scrapie in sheep, BSE in cattle and Cruetzfeldt-Jakob disease in humans.

Important Note

This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.

Background Descriptions

The function of PrP is still under debate. May play a role in neuronal development and synaptic plasticity. May be required for neuronal myelin sheath maintenance. May play a role in iron uptake and iron homeostasis (By similarity). Isoform 2 may act as a growth suppressor by arresting the cell cycle at the G0/G1 phase. Soluble oligomers are toxic to cultured neuroblastoma cells and induce apoptosis (in vitro).

Additional Information

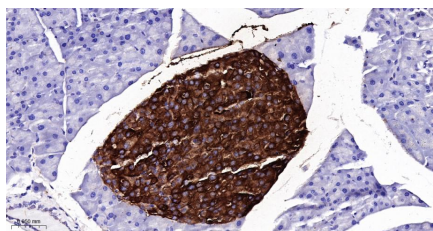
Dilution	WB=1:500-2000,IHC-P=1:50-200,IHC-F=1:50-200,IF=1:50-200,ICC/IF=1:50-200, Flow-Cyt=1:50-100
Storage	Store at -20 °C for one year. Avoid repeated freeze/thaw cycles. When reconstituted in sterile pH 7.4 0.01M PBS or diluent of antibody the antibody is stable for at least two weeks at 2-4 °C.

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Images

Paraformaldehyde-fixed, paraffin embedded Mouse



Pancreas; Antigen retrieval by boiling in sodium citrate buffer (pH6.0) for 15 min; The section was incubated with PRNP Monoclonal Antibody, Unconjugated (AP94218) at 1:200 overnight at 4°C, followed by conjugation to the AP94218-HRP and DAB (C-0010) staining.

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