

Prion protein PrP/CD230 Polyclonal Antibody
Purified Rabbit Polyclonal Antibody (Pab)
Catalog # AP54624**Specification****Prion protein PrP/CD230 Polyclonal Antibody - Product Information**

Application	IHC-P, IHC-F, IF, ICC, E
Primary Accession	P04156
Reactivity	Rat, Bovine
Host	Rabbit
Clonality	Polyclonal
Calculated MW	25 KDa
Physical State	Liquid
Immunogen	KLH conjugated synthetic peptide derived from human PRNP
Epitope Specificity	23-120/253
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.
SIMILARITY	Belongs to the prion family.
SUBUNIT	Monomer and homodimer. Has a tendency to aggregate into amyloid fibrils containing a cross-beta spine, formed by a steric zipper of superposed beta-strands. Soluble oligomers may represent an intermediate stage on the path to fibril formation. Copper binding may promote oligomerization. Interacts with GRB2, APP, ERI3/PRNP and SYN1. Mislocalized cytosolically exposed PrP interacts with MGRN1; this interaction alters MGRN1 subcellular location and causes lysosomal enlargement (By similarity). Interacts with KIAA1191.
Post-translational modifications	The glycosylation pattern (the amount of mono-, di- and non-glycosylated forms or glycoforms) seems to differ in normal and CJD prion. Isoform 2 is sumoylated by SUMO1.
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 (PrPc) is converted to the disease form, PrPSc, through alterations in the protein folding conformations. PrPc is constitutively expressed in normal adult brain and is sensitive to proteinase K digestion, while the altered PrPSc 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 PrPc with PrPSc both in vitro and in vivo produces PrPc that is resistant to protease degradation. Infectious PrPSc 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.

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

Important Note

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).

Prion protein PrP/CD230 Polyclonal Antibody - Additional Information

Gene ID 5621

Other Names

Major prion protein, PrP, ASCR, PrP27-30, PrP33-35C, CD230, PRNP, ALTPRP, PRIP, PRP

Dilution

IHC-P ~ ~ N/A
IHC-F ~ ~ N/A
IF ~ ~ 1:50 ~ 200
ICC ~ ~ N/A
E ~ ~ N/A

Format

0.01M TBS(pH7.4), 0.09% (W/V) sodium azide and 50% Glyce

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.

Prion protein PrP/CD230 Polyclonal Antibody - Protein Information

Name PRNP

Synonyms ALTPRP, PRIP, PRP

Function

Its primary physiological function is unclear. May play a role in neuronal development and synaptic plasticity. May be required for neuronal myelin sheath maintenance. May promote myelin homeostasis through acting as an agonist for ADGRG6 receptor. May play a role in iron uptake and iron homeostasis. Soluble oligomers are toxic to cultured neuroblastoma cells and induce apoptosis (in vitro) (By similarity). Association with GPC1 (via its heparan sulfate chains) targets PRNP to lipid rafts. Also provides Cu(2+) or Zn(2+) for the ascorbate-mediated GPC1 deaminase degradation of its heparan sulfate side chains (By similarity).

Cellular Location

Cell membrane; Lipid-anchor, GPI-anchor. Golgi apparatus {ECO:0000250|UniProtKB:P04925}. Note=Targeted to lipid rafts via association with the heparan sulfate chains of GPC1. Colocates, in the presence of Cu(2+), to vesicles in para- and perinuclear regions, where both proteins undergo internalization. Heparin displaces PRNP from lipid rafts and promotes endocytosis.

Prion protein PrP/CD230 Polyclonal Antibody - Protocols

Provided below are standard protocols that you may find useful for product applications.

- [Western Blot](#)
- [Blocking Peptides](#)
- [Dot Blot](#)
- [Immunohistochemistry](#)

- [Immunofluorescence](#)
- [Immunoprecipitation](#)
- [Flow Cytometry](#)
- [Cell Culture](#)

Prion protein PrP/CD230 Polyclonal Antibody - Images