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Rabbit Anti-Prion protein PrP antibody

SL4728R

Product Name:	Prion protein PrP
Chinese Name:	朊病毒/感染性蛋白抗体
Alias:	AltPrP; ASCR; atal familial insomnia; CD230; CD230 antigen; CJD; Creutzfeld Jakob disease; Gerstmann-Strausler-Scheinker syndrome; GSS; KURU; Major prion protein; MGC26679; p27 30; PRIO_HUMAN; Prion protein; Prion related protein; PRIP; Prni; Prnp; PrP; PrP27 30; PrP27-30; PrP33 35C; PrP33-35C; PrPC; PrPSc; Sinc.
Organism Species:	Rabbit
Clonality:	Polyclonal
React Species:	Human,Mouse,Rat,Cow,Horse,Sheep,
Applications:	ELISA=1:500-1000IHC-P=1:400-800IHC-F=1:400-800ICC=1:100-500IF=1:100-500（Paraffin sections need antigen repair） not yet tested in other applications. optimal dilutions/concentrations should be determined by the end user.
Molecular weight:	23kDa
Cellular localization:	The nucleuscytoplasmicThe cell membrane
Form:	Lyophilized or Liquid
Concentration:	1mg/ml
immunogen:	KLH conjugated synthetic peptide derived from human Major prion protein:51-150/253
Lsotype:	IgG
Purification:	affinity purified by Protein A
Storage Buffer:	0.01M TBS(pH7.4) with 1% BSA, 0.03% Proclin300 and 50% Glycerol.
Storage:	Store at -20 °C for one year. Avoid repeated freeze/thaw cycles. The lyophilized antibody is stable at room temperature for at least one month and for greater than a year when kept at -20°C. 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.
PubMed:	PubMed
Product Detail:	The protein encoded by this gene is a membrane glycosylphosphatidylinositol-anchored glycoprotein that tends to aggregate into rod-like structures. The encoded protein contains a highly unstable region of five tandem octapeptide repeats. This gene is found on chromosome 20, approximately 20 kbp upstream of a gene which encodes a

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	biochemically and structurally similar protein to the one encoded by this gene. Mutations in the repeat region as well as elsewhere in this gene have been associated with Creutzfeldt-Jakob disease, fatal familial insomnia, Gerstmann-Straussler disease, Huntington disease-like 1, and kuru. An overlapping open reading frame has been found for this gene that encodes a smaller, structurally unrelated protein, AltPrp. Alternative splicing results in multiple transcript variants. [provided by RefSeq, Oct 2012]. **Function:** 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. Soluble oligomers are toxic to cultured neuroblastoma cells and induce apoptosis (in vitro). Association with GPC1 (via its heparin 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. **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/PRNPIP and SYN1. Mislocalized cytosolically exposed PrP interacts with MGRN1; this interaction alters MGRN1 subcellular location and causes lysosomal enlargement. Interacts with KIAA1191. **Subcellular Location:** Cell membrane; Lipid-anchor, GPI-anchor. Golgi apparatus. 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. Isoform 2: Cytoplasm. Nucleus. Note=Accumulates outside the secretory route in the cytoplasm, from where it relocates to the nucleus. **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 with 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

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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 annimal 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.

Similarity:
Belongs to the prion family.

SWISS:
P04156

Gene ID:

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	5621 **Database links:** Entrez Gene: 5621Human Entrez Gene: 19122Mouse Entrez Gene: 24686Rat Omim: 176640Human SwissProt: P04156Human SwissProt: P04925Mouse SwissProt: P13852Rat Unigene: 472010Human Unigene: 610285Human Unigene: 727471Human Unigene: 648Mouse Unigene: 3936Rat **Important Note:** This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.

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