

Product datasheet

Anti-Prion protein PrP antibody ab703

[3 References](#) [1 Image](#)

Overview

Product name	Anti-Prion protein PrP antibody
Description	Rabbit polyclonal to Prion protein PrP
Host species	Rabbit
Specificity	Recognises natural PrPc and conformationally changed PrPc (PrPsc). Recognises recombinant protein , ab753 , in western blot and ELISA.
Tested applications	Suitable for: WB
Species reactivity	Reacts with: Mouse
Immunogen	Recombinant full length protein corresponding to Cow Prion protein PrP aa 1-250. Database link: P10279
General notes	<p>The Life Science industry has been in the grips of a reproducibility crisis for a number of years. Abcam is leading the way in addressing this with our range of recombinant monoclonal antibodies and knockout edited cell lines for gold-standard validation. Please check that this product meets your needs before purchasing.</p> <p>If you have any questions, special requirements or concerns, please send us an inquiry and/or contact our Support team ahead of purchase. Recommended alternatives for this product can be found below, along with publications, customer reviews and Q&As</p>

Properties

Form	Liquid
Storage instructions	Shipped at 4°C. Store at +4°C short term (1-2 weeks). Upon delivery aliquot. Store at -20°C or -80°C. Avoid freeze / thaw cycle.
Storage buffer	Preservative: 0.02% Sodium azide Constituent: Whole serum
Purity	Whole antiserum
Clonality	Polyclonal
Isotype	IgG

Applications

The Abpromise guarantee

Our [Abpromise guarantee](#) covers the use of ab703 in the following tested applications.

The application notes include recommended starting dilutions; optimal dilutions/concentrations should be determined by the end user.

Application	Abreviews	Notes
WB		1/2000.

Target

Function

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).

Involvement in 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.

Sequence similarities

Belongs to the prion family.

Domain

The normal, monomeric form has a mainly alpha-helical structure. The disease-associated, protease-resistant form forms amyloid fibrils containing a cross-beta spine, formed by a steric zipper of superposed beta-strands. Disease mutations may favor intermolecular contacts via short beta strands, and may thereby trigger oligomerization.

Contains an N-terminal region composed of octamer repeats. At low copper concentrations, the sidechains of His residues from three or four repeats contribute to the binding of a single copper ion. Alternatively, a copper ion can be bound by interaction with the sidechain and backbone amide nitrogen of a single His residue. The observed copper binding stoichiometry suggests that two repeat regions cooperate to stabilize the binding of a single copper ion. At higher copper concentrations, each octamer can bind one copper ion by interactions with the His sidechain and Gly backbone atoms. A mixture of binding types may occur, especially in the case of octamer repeat expansion. Copper binding may stabilize the conformation of this region and may promote oligomerization.

Post-translational modifications

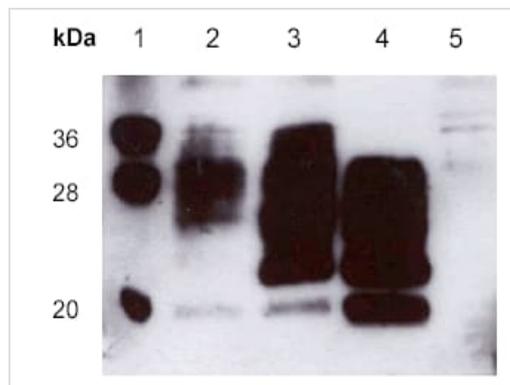
The glycosylation pattern (the amount of mono-, di- and non-glycosylated forms or glycoforms) seems to differ in normal and CJD prion.

Isoform 2 is sumoylated by SUMO1.

Cellular localization

Cell membrane. Golgi apparatus and Cytoplasm. Nucleus. Accumulates outside the secretory route in the cytoplasm, from where it relocates to the nucleus.

Images



Western blot - Anti-Prion protein PrP antibody (ab703)

This image is courtesy of Dr. M. Glatzel, Institut of Neuropathology, University Hospital of Zürich, Switzerland

Lane 1: MW standard

Lane 2: wild-type mice (PrPc)

Lane 3: CDI/PrPsc (PK-)

Lane 4: CDI/PrPsc (PK+),

Lane 5: PrP 0/0 mice

CDI = clinically ill CD-1 mice, PK = digested with Proteinase K.

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