abcam

## Product datasheet

# Recombinant Cow Prion protein PrP (mutated F209S) ab90587

### 製品の詳細

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| **製品名** | Recombinant Cow Prion protein PrP (mutated F209S) |
| **精製度** | > 95 % SDS-PAGE. |
| **発現系** | Escherichia coli |
| **タンパク質長** | Full length protein |
| **Animal free** | No |
| **由来** | Recombinant |
| **生物種** | Cow |

### 特性

Our **Abpromise guarantee** covers the use of **ab90587** in the following tested applications.

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

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| **アプリケーション** | SDS-PAGE<br>Functional Studies |
| **製品の状態** | Lyophilized |

### 前処理および保存

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| **保存方法および安定性** | Shipped on dry ice. Upon delivery aliquot and store at -80ºC. Avoid freeze / thaw cycles.<br>pH: 6.70<br>Constituent: 0.158% Tris HCl |
| **再構成** | The protein may be reconstituted in detergent-containing buffers e.g. TX-100 (0.5%) or under mild denaturing conditions (e.g. 1.5 M guanidine-HCl or urea). |

### 関連情報

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| **機能** | 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).

**関連疾患**

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 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.

**配列類似性**

Belongs to the prion family.

**ドメイン**

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

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| | 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. |
| **翻訳後修飾** | The glycosylation pattern (the amount of mono-, di- and non-glycosylated forms or glycoforms) seems to differ in normal and CJD prion.<br>Isoform 2 is sumoylated by SUMO1. |
| **細胞内局在** | Cell membrane. Golgi apparatus and Cytoplasm. Nucleus. Accumulates outside the secretory route in the cytoplasm, from where it relocates to the nucleus. |

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