abcam

Product datasheet

Recombinant Human Prion protein PrP (denatured) ab140567

1 图像

描述

产品名称	重组人Prion蛋白PrP (denatured)
纯度	> 90 % SDS-PAGE.
表达系统	Escherichia coli
Accession	P04156
蛋白长度	Full length protein
无动物成分	No
性质	Recombinant
种属	Human
序列	MGSSHHHHHH SSGLVPRGSH MKKRPKPGGW NTGGSRYPGQ GSPGGNRYPP QGGGGWGQPH GGGWGQPHGG GWGQPHGGGW GQPHGGGWGQ GGGTHSQWNK PSKPKTNMKH MAGAAAAGAV VGGLGGYVLG SAMSRPIIHF GSDYEDRYYR ENMHRYPNQV YYRPMDEYSN QNNFVHDCVN ITIKQHTVTT TTKGENFTET DVKMMERVVE QMCITQYERE SQAYYQRGS
预测分子量	25 kDa including tags
氨基酸	23 to 230
标签	His tag N-Terminus

技术指标

Our **Abpromise guarantee** covers the use of **ab140567** in the following tested applications.

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

应用	SDS-PAGE
形式	Liquid

制备和贮存

稳定性和存储	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.

pH: 8.00
Constituents: 6.01% Urea, 0.32% Tris HCl, 10% Glycerol

常规信息

功能

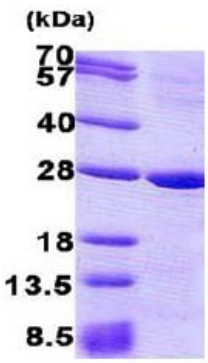
The function of PrP is still under debate. May play a role in neuronal development and synaptic plasticity. May be required for neuronal myelin sheath maintenance. May play a role in iron uptake and iron homeostasis (By similarity). Isoform 2 may act as a growth suppressor by arresting the cell cycle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

疾病相关

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.
序列相似性	Belongs to the prion family.
结构域	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.
翻译后修饰	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.
细胞定位	Cell membrane. Golgi apparatus and Cytoplasm. Nucleus. Accumulates outside the secretory route in the cytoplasm, from where it relocates to the nucleus.

图片



SDS-PAGE - Recombinant Human Prion protein PrP (denatured) (ab140567)

15% SDS PAGE analysis of ab140567 (3 µg).

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