

## Product datasheet

# Recombinant Human Huntingtin protein (Tagged) ab112300

[1 Image](#)

### Description

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<b>Product name</b>	Recombinant Human Huntingtin protein (Tagged)
<b>Biological activity</b>	useful for Antibody Production and Protein Array
<b>Expression system</b>	Wheat germ
<b>Accession</b>	<b><u>P42858</u></b>
<b>Protein length</b>	Protein fragment
<b>Animal free</b>	No
<b>Nature</b>	Recombinant
<b>Species</b>	Human
<b>Sequence</b>	AVAEELHRPKKELSATKKDRVNHCLTICENVAQSVRNS PEFQKLLGIA MELFLLCSDDAESDVRMVADECLNKVIKALMDSNLPRLQ LELYKEIKKNG APRSLRAALW
<b>Predicted molecular weight</b>	38 kDa including tags
<b>Amino acids</b>	81 to 190
<b>Tags</b>	GST tag N-Terminus
<b>Additional sequence information</b>	NP_002102

### Specifications

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Our **Abpromise guarantee** covers the use of **ab112300** in the following tested applications.

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

<b>Applications</b>	ELISA SDS-PAGE Western blot
<b>Form</b>	Liquid
<b>Additional notes</b>	Useful for Antibody Production and Protein Array

### Preparation and Storage

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## Stability and Storage

Shipped on dry ice. Upon delivery aliquot and store at -80°C. Avoid freeze / thaw cycles.

pH: 8.00

Constituents: 0.31% Glutathione, 0.79% Tris HCl

Glutathione is reduced

## General Info

### Function

May play a role in microtubule-mediated transport or vesicle function.

### Tissue specificity

Expressed in the brain cortex (at protein level). Widely expressed with the highest level of expression in the brain (nerve fibers, varicosities, and nerve endings). In the brain, the regions where it can be mainly found are the cerebellar cortex, the neocortex, the striatum, and the hippocampal formation.

### Involvement in disease

Defects in HTT are the cause of Huntington disease (HD) [MIM:143100]. HD is an autosomal dominant neurodegenerative disorder characterized by involuntary movements (chorea), general motor impairment, psychiatric disorders and dementia. Onset of the disease occurs usually in the third or fourth decade of life and symptoms progressively worsen leading to death in 10 to 20 years. Onset and clinical course depend on the degree of poly-Gln repeat expansion, longer expansions resulting in earlier onset and more severe clinical manifestations. HD affects 1 in 10,000 individuals of European origin. Neuropathology of Huntington disease displays a distinctive pattern with loss of neurons, especially in the caudate and putamen (striatum).

### Sequence similarities

Belongs to the huntingtin family.

Contains 10 HEAT repeats.

### Domain

The N-terminal Gln-rich and Pro-rich domain has great conformational flexibility and is likely to exist in a fluctuating equilibrium of alpha-helical, random coil, and extended conformations.

### Post-translational modifications

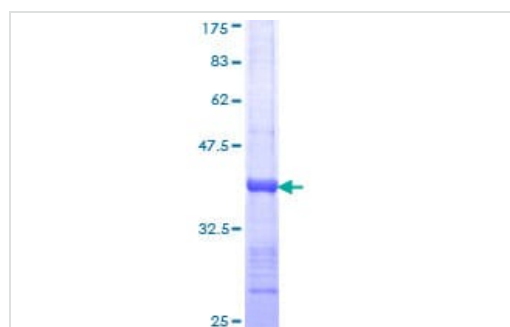
Cleaved by apopain downstream of the polyglutamine stretch. The resulting N-terminal fragment is cytotoxic and provokes apoptosis.

Forms with expanded polyglutamine expansion are specifically ubiquitinated by SYVN1, which promotes their proteasomal degradation.

### Cellular localization

Cytoplasm. Nucleus. The mutant Huntingtin protein colocalizes with AKAP8L in the nuclear matrix of Huntington's disease neurons.

## Images



ab112300 analysed on a 12.5% SDS-PAGE stained with Coomassie Blue.

SDS-PAGE - Recombinant Human Huntingtin protein (ab112300)

**Please note:** All products are "FOR RESEARCH USE ONLY. NOT FOR USE IN DIAGNOSTIC PROCEDURES"

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