abcam

Product datasheet

Recombinant Human Ataxin 3 protein ab86706

1 Image

Description

Product name Recombinant Human Ataxin 3 protein

Purity > 90 % SDS-PAGE.

ab86706 is purified using conventional chromatography techniques. Endotoxin Level: < 1.0 EU

per 1 µg of protein (determined by LAL method).

Expression system Escherichia coli

Accession P54252

Protein length Full length protein

Animal free No

Nature Recombinant

Species Human

Sequence MESIFHEKQE GSLCAQHCLN NLLQGEYFSP

VELSSIAHQL DEEERMRMAE GGVTSEDYRT FLQQPSGNMD DSGFFSIQVI SNALKVWGLE

LILFNSPEYQ RLRIDPINER SFICNYKEHW FTVRKLGKQW FNLNSLLTGP ELISDTYLAL FLAQLQQEGY SIFVVKGDLP

DCEADQLLQM IRVQQMHRPK LIGEELAQLK
EQRVHKTDLE RVLEANDGSG MLDEDEEDLQ
RALALSRQEI DMEDEEADLR RAIQLSMQGS
SRNISQDMTQ TSGTNLTSEE LRKRREAYFE
KQQQKQQQQQ QQQQQQQQQQQQQQDLSGQ
SSHPCERPAT SSGALGSDLG DAMSEEDMLQ

AAVTMSLETV RNDLKTEGKK

Predicted molecular weight 42 kDa

Amino acids 1 to 370

Specifications

Our Abpromise guarantee covers the use of ab86706 in the following tested applications.

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

Applications SDS-PAGE

Form Liquid

1

Preparation and Storage

Stability and Storage

Shipped at 4°C. Upon delivery aliquot and store at -20°C or -80°C. Avoid repeated freeze / thaw cycles.

pH: 7.50

Constituents: 0.0308% DTT, 0.316% Tris HCI, 10% Glycerol (glycerin, glycerine), 0.29% Sodium

chloride

General Info

Function

Interacts with key regulators (CBP, p300 and PCAF) of transcription and represses transcription. Acts as a histone-binding protein that regulates transcription. Acts as a deubiquitinating enzyme.

Tissue specificity

Ubiquitous.

Involvement in disease

Defects in ATXN3 are the cause of spinocerebellar ataxia type 3 (SCA3) [MIM:109150]; also known as Machado-Joseph disease (MJD). Spinocerebellar ataxia is a clinically and genetically heterogeneous group of cerebellar disorders. Patients show progressive incoordination of gait and often poor coordination of hands, speech and eye movements, due to degeneration of the cerebellum with variable involvement of the brainstem and spinal cord. SCA3 belongs to the autosomal dominant cerebellar ataxias type I (ADCA I) which are characterized by cerebellar ataxia in combination with additional clinical features like optic atrophy, ophthalmoplegia, bulbar and extrapyramidal signs, peripheral neuropathy and dementia. The molecular defect in SCA3 is the a CAG repeat expansion in ATXN3 coding region. Longer expansions result in earlier onset and more severe clinical manifestations of the disease.

Sequence similarities

Contains 1 Josephin domain.

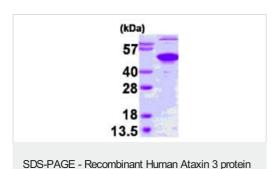
Contains 3 UIM (ubiquitin-interacting motif) repeats.

Cellular localization

Nucleus matrix. Predominantly nuclear, but not exclusively, inner nuclear matrix.

Images

(ab86706)



15% SDS-PAGE showing ab86706 at approximately 42.4kDa ($3\mu g$).

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