



Atrophin-1 Monoclonal Antibody

Catalog No	YP-mAb-12687
Isotype	IgG
Reactivity	Human;Mouse;Rat
Applications	WB
Gene Name	ATN1
Immunogen	The antiserum was produced against synthesized peptide derived from human ATN1. AA range:81-130
Specificity	Atrophin-1 Monoclonal Antibody detects endogenous levels of Atrophin-1 protein.
Formulation	Liquid in PBS containing 50% glycerol, 0.5% BSA and 0.02% sodium azide.
Source	Monoclonal, Mouse,IgG
Purification	The antibody was affinity-purified from mouse antiserum by affinity-chromatography using epitope-specific immunogen.
Dilution	WB 1:500-1:2000
Concentration	1 mg/ml
Purity	≥90%
Storage Stability	-20°C/1 year
Synonyms	ATN1;atrophin 1;B37;D12S755E;DRPLA;HRS;NOD;Atrophin-1;Dentatorubral-pallidoluysian atrophy protein
Observed Band	130kD
Calculated Molecular Weight	130kD
Cell Pathway	Nucleus. Cytoplasm, perinuclear region. Cell junction . Shuttles between nucleus and cytoplasm. Colocalizes with FAT1 in the perinuclear area, at cell-cell junctions and leading edges of cells (By similarity). Colocalizes with MTG8 in discrete nuclear dots. Proteolytic fragment F1 appears to remain in nucleus. Fragment F2 is exported into the cytoplasm. Fragment F2 from mutant sequences with longer poly-Gln (polyQ) tracts are additionally located to the cytoplasmic membrane and to certain organelles. .
Tissue Specificity	Widely expressed in various tissues including heart, lung, kidney, ovary, testis, prostate, placenta, skeletal Low levels in the liver, thymus and leukocytes. In the adult brain, broadly expressed in amygdala, caudate nucleus, corpus callosum, hippocampus, hypothalamus, substantia nigra, subthalamic nucleus, and thalamus. High levels in fetal tissues, especially brain.



Function

disease: Defects in ATN1 are the cause of dentatorubral-pallidoluysian atrophy (DRPLA) [MIM:125370]. DRPLA is an autosomal dominant neurodegenerative disorder characterized by a loss of neurons in the dentate nucleus, rubrum, globus pallidus and Luys' body. Clinical features are myoclonus epilepsy, dementia, and cerebellar ataxia. Onset of the disease occurs usually in the second decade of life and death in the fourth. polymorphism: The poly-Gln region of ATN1 is highly polymorphic (7 to 23 repeats) in the normal population and is expanded to about 49-75 repeats in DRPLA and HRS patients. Longer expansions result in earlier onset and more severe clinical manifestations of the disease. subunit: Interacts with BAIAP2, WWP1, WWP2, WWP3 and RERE. tissue specificity: Relatively high levels in the brain, ovary, testis and prostate. Lower levels in the liver, thymus and leukocytes.

Background

Dentatorubral pallidoluysian atrophy (DRPLA) is a rare neurodegenerative disorder characterized by cerebellar ataxia, myoclonic epilepsy, choreoathetosis, and dementia. The disorder is related to the expansion from 7-35 copies to 49-93 copies of a trinucleotide repeat (CAG/CAA) within this gene. The encoded protein includes a serine repeat and a region of alternating acidic and basic amino acids, as well as the variable glutamine repeat. Alternative splicing results in two transcripts variants that encode the same protein. [provided by RefSeq, Jul 2016],

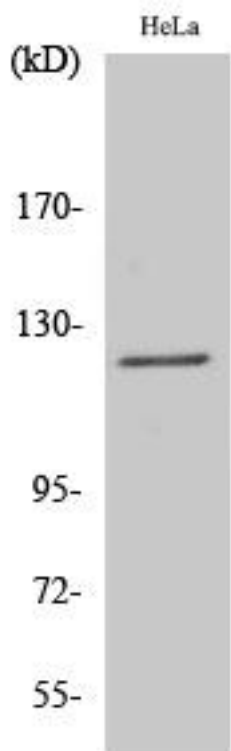
matters needing attention

Avoid repeated freezing and thawing!

Usage suggestions

This product can be used in immunological reaction related experiments. For more information, please consult technical personnel.

Products Images



Western Blot analysis of various cells using Atrophin-1 Monoclonal Antibody