Product name:	Atrophin-1 Rabbit Polyclonal Antibody
Cat number:	ABN07356
Conjugate:	Unconjugated
Size:	100µL
Clone:	Polyclonal
Concentration:	1mg/ml
Host:	Rabbit
Isotype:	IgG
Immunogen:	The antiserum was produced against synthesized peptide derived from human ATN1. AA range:81-130
Reactivity:	Human,Mouse,Rat
Applications:	WB 1:500-1:2000,ELISA 1:5000-1:10000
Molecular Weight:	130kDa
Purification:	Affinity purification
Form:	Liquid
Buffer:	Liquid in PBS containing 50% glycerol, 0.5% BSA and 0.02% New type preservative N.
Storage:	Store at 4°C short term. Aliquot and store at -20°C for 12 months. Avoid freeze/thaw cycles.

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],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, glogus 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.,