SANTA CRUZ BIOTECHNOLOGY, INC.

# atrophin-1 (C-18): sc-10304

## BACKGROUND

Dentatorubral-pallidoluysian atrophy protein, also designated Atrophin-1, interacts with several other proteins, including RERE, BAIAP2 and WWP1-3. It is highly expressed in ovary, testis, brain and prostate, but can also be detected in thymus, liver, and leukocytes. Defects in the gene encoding for the Atrophin protein, ATN1, can cause dentatorubral-pallidoluysian atrophy (DRPLA) or Haw River syndrome (HRS). Both disorders are dominant neurodegenerative disorders caused by an increase in the number of polyglutamine (Gln) repeats in the ATN1 gene (7-23 repeats in the normal population, 49-75 in patients affected by DRPLA or HRS). More repeats corresponds to earlier onset and more severe clinical manifestations of the diseases. DRPLA is characterized by a loss of neurons in the dentate nucleus, rubrum, globus pallidus and Luys' body, often resulting in dementia, epilepsy and cerebellar ataxia. HRS is characterized by the degeneration of multiple systems, resembling that of DRPLA or Huntington's disease.

## REFERENCES


1. Nagafuchi, S., et al. 1994. Structure and expression of the gene responsible for the triplet repeat disorder, dentatorubral and pallidoluysian atrophy (DRPLA). Nat. Gene 8: 177-182.
2. Yazawa, I., et al. 1995. Abnormal gene product identified in hereditary dentatorubral-pallidoluysian atrophy (DRPLA) brain. Nat. Genet. 10: 99-103.
3. Miyashita, T., et al. 1997. Dentatorubral pallidoluysian atrophy (DRPLA) protein is cleaved by caspase-3 during apoptosis. J. Biol. Chem. 272: 29238-29242.
4. Wood, J.D., et al. 1998. Atrophin-1, the DRPLA gene product, interacts with two families of WW domain-containing proteins. Mol. Cell. Neurosci. 11: 149-160.
5. Kanazawa, I. 1998. Dentatorubral-pallidoluysian atrophy or Naito-Oyanagi disease. Neurogenetics 2: 1-17.
6. Schilling, G., et al. 1999. Nuclear accumulation of truncated atrophin-1 fragments in a transgenic mouse model of DRPLA. Neuron 24: 275-286.


## SOURCE

atrophin-1 (C-18) is an affinity purified goat polyclonal antibody raised against a peptide mapping near the C-terminus of atrophin-1 of human origin.

## PRODUCT

Each vial contains 200 µg IgG in 1.0 ml of PBS with < 0.1% sodium azide and 0.1% gelatin.

Blocking peptide available for competition studies, sc-10304 P, (100 µg peptide in 0.5 ml PBS containing < 0.1% sodium azide and 0.2% BSA).

## STORAGE

Store at 4° C, **DO NOT FREEZE**. Stable for one year from the date of shipment. Non-hazardous. No MSDS required.

## RESEARCH USE

For research use only, not for use in diagnostic procedures.

## APPLICATIONS

atrophin-1 (C-18) is recommended for detection of atrophin-1 of mouse, rat and human origin by Western Blotting (starting dilution 1:200, dilution range 1:100-1:1000), immunofluorescence (starting dilution 1:50, dilution range 1:50-1:500) and solid phase ELISA (starting dilution 1:30, dilution range 1:30-1:3000).

Suitable for use as control antibody for atrophin-1 siRNA (h): sc-29765, atrophin-1 siRNA (m): sc-29766, atrophin-1 shRNA Plasmid (h): sc-29765-SH, atrophin-1 shRNA Plasmid (m): sc-29766-SH, atrophin-1 shRNA (h) Lentiviral Particles: sc-29765-V and atrophin-1 shRNA (m) Lentiviral Particles: sc-29766-V.

Molecular Weight of atrophin-1: 150-180 kDa.

Positive Controls: EOC 20, mouse placenta extract or mouse brain extract: sc-2253.

## RECOMMENDED SECONDARY REAGENTS

To ensure optimal results, the following support (secondary) reagents are recommended: 1) Western Blotting: use donkey anti-goat IgG-HRP: sc-2020 (dilution range: 1:2000-1:100,000) or Cruz Marker™ compatible donkey anti-goat IgG-HRP: sc-2033 (dilution range: 1:2000-1:5000), Cruz Marker™ Molecular Weight Standards: sc-2035, TBS Blotto A Blocking Reagent: sc-2333 and Western Blotting Luminol Reagent: sc-2048. 2) Immunofluorescence: use donkey anti-goat IgG-FITC: sc-2024 (dilution range: 1:100-1:400) or donkey anti-goat IgG-TR: sc-2783 (dilution range: 1:100-1:400) with UltraCruz™ Mounting Medium: sc-24941.

## PROTOCOLS

See our web site at www.scbt.com or our catalog for detailed protocols and support products.