

atrophin-1 (H-70): sc-28191

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 ATN1, the gene encoding for the atrophin protein, 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. Genet.* 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. Margolis, R.L., et al. 1996. DRPLA gene (atrophin-1) sequence and mRNA expression in human brain. *Brain Res. Mol. Brain Res.* 36: 219-226.
4. Miyashita, T., et al. 1997. Dentatorubral pallidoluysian atrophy (DRPLA) protein is cleaved by caspase-3 during apoptosis. *J. Biol. Chem.* 272: 29238-29242.

CHROMOSOMAL LOCATION

Genetic locus: ATN1 (human) mapping to 12p13.31; Atn1 (mouse) mapping to 6 F2.

SOURCE

atrophin-1 (H-70) is a rabbit polyclonal antibody raised against amino acids 19-88 mapping near the N-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.

STORAGE

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

PROTOCOLS

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

APPLICATIONS

atrophin-1 (H-70) 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), immunoprecipitation [1-2 µg per 100-500 µg of total protein (1 ml of cell lysate)], 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).

atrophin-1 (H-70) is also recommended for detection of atrophin-1 in additional species, including equine and porcine.

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.

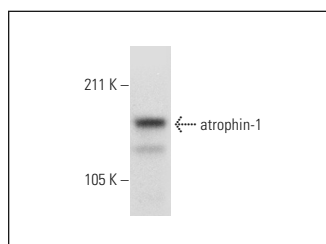
Molecular Weight of atrophin-1: 150-180 kDa.

Positive Controls: EOC 20 whole cell lysate: sc-364187, T98G cell lysate: sc-2294 or mouse brain extract: sc-2253.

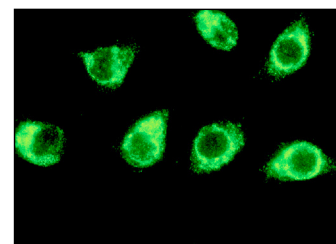
RECOMMENDED SECONDARY REAGENTS

To ensure optimal results, the following support (secondary) reagents are recommended: 1) Western Blotting: use goat anti-rabbit IgG-HRP: sc-2004 (dilution range: 1:2000-1:100,000) or Cruz Marker™ compatible goat anti-rabbit IgG-HRP: sc-2030 (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) Immunoprecipitation: use Protein A/G PLUS-Agarose: sc-2003 (0.5 ml agarose/2.0 ml). 3) Immunofluorescence: use goat anti-rabbit IgG-FITC: sc-2012 (dilution range: 1:100-1:400) or goat anti-rabbit IgG-TR: sc-2780 (dilution range: 1:100-1:400) with UltraCruz™ Mounting Medium: sc-24941.

DATA



atrophin-1 (H-70): sc-28191. Western blot analysis of atrophin-1 expression in T98G whole cell lysate.



atrophin-1 (H-70): sc-28191. Immunofluorescence staining of methanol-fixed EOC 20 cells showing cytoplasmic localization.

RESEARCH USE

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