

atrophin-1 (C1): sc-517594

BACKGROUND

Dentatorubral-pallidolusian 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-pallidolusian 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

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- Miyashita, T., et al. 1997. Dentatorubral pallidolusian atrophy (DRPLA) protein is cleaved by caspase-3 during apoptosis. *J. Biol. Chem.* 272: 29238-29242.
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CHROMOSOMAL LOCATION

Genetic locus: ATN1 (human) mapping to 12p13.31.

SOURCE

atrophin-1 (C1) is a mouse monoclonal antibody raised against recombinant atrophin-1 of human origin.

PRODUCT

Each vial contains 100 µg IgG_{2b} kappa light chain in 1.0 ml of PBS with < 0.1% sodium azide, 0.1% gelatin and 5% glycerol.

APPLICATIONS

atrophin-1 (C1) is recommended for detection of atrophin-1 of 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 immunohistochemistry (including paraffin-embedded sections) (starting dilution 1:50, dilution range 1:50-1:500).

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

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

RECOMMENDED SUPPORT REAGENTS

To ensure optimal results, the following support reagents are recommended: 1) Western Blotting: use m-IgGκ BP-HRP: sc-516102 or m-IgGκ BP-HRP (Cruz Marker): sc-516102-CM (dilution range: 1:1000-1:10000), Cruz Marker™ Molecular Weight Standards: sc-2035, UltraCruz® Blocking Reagent: sc-516214 and Western Blotting Luminol Reagent: sc-2048. 2) Immunofluorescence: use m-IgGκ BP-FITC: sc-516140 or m-IgGκ BP-PE: sc-516141 (dilution range: 1:50-1:200) with UltraCruz® Mounting Medium: sc-24941 or UltraCruz® Hard-set Mounting Medium: sc-359850. 3) Immunohistochemistry: use m-IgGκ BP-HRP: sc-516102 with DAB, 50X: sc-24982 and Immunohistomount: sc-45086, or Organo/Limonene Mount: sc-45087.

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.

PROTOCOLS

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