

Ataxin-2 (C-18): sc-18478

BACKGROUND

Autosomal dominant cerebellar ataxias are a group of neurodegenerative disorders caused by unstable CAG repeat expansions encoding polyglutamine tracts. Proteins with long polyglutamine tracts have an increased tendency to aggregate, often forming ubiquitinated intranuclear inclusion bodies. Ataxin-2, the gene product of the human spinocerebellar ataxia type 2 (SCA2) gene, is a basic protein with two domains (Sm1 and Sm2) implicated in RNA splicing and protein interaction. Ataxin-2 interacts with a putative RNA-binding protein Ataxin-2-binding-protein 1 (A2BP1), which is expressed in muscle and brain. Ataxin-2 is ubiquitously expressed with highest levels in the cytoplasm of Purkinje cells. Both A2BP1 and Ataxin-2 are localized to the *trans*-Golgi network. Mice expressing Ataxin-2 with polyglutamine show progressive functional deficits accompanied by loss of Purkinje cell dendritic arbor and eventually loss of Purkinje cells. In conclusion, expansion of Ataxin-2 results in spinocerebellar ataxia type 2, which affects the cerebellum and other areas of the central nervous system.

REFERENCES

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- Huynh, D.P., et al. 2000. Nuclear localization or inclusion body formation of ataxin-2 are not necessary for SCA2 pathogenesis in mouse or human. *Nat. Genet.* 26: 44-50.
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- Shibata, H., et al. 2000. A novel protein with RNA-binding motifs interacts with Ataxin-2. *Hum. Mol. Genet.* 9: 1303-1313.

CHROMOSOMAL LOCATION

Genetic locus: ATXN2 (human) mapping to 12q24.12; Atxn2 (mouse) mapping to 5 F.

SOURCE

Ataxin-2 (C-18) is an affinity purified goat polyclonal antibody raised against a peptide mapping at the C-terminus of Ataxin-2 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-18478 P, (100 µg peptide in 0.5 ml PBS containing < 0.1% sodium azide and 0.2% BSA).

APPLICATIONS

Ataxin-2 (C-18) is recommended for detection of Ataxin-2 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), immunohistochemistry (including paraffin-embedded sections) (starting dilution 1:50, dilution range 1:50-1:500) and solid phase ELISA (starting dilution 1:30, dilution range 1:30-1:3000).

Ataxin-2 (C-18) is also recommended for detection of Ataxin-2 in additional species, including equine, canine, bovine, porcine and avian.

Suitable for use as control antibody for Ataxin-2 siRNA (h): sc-40356, Ataxin-2 siRNA (m): sc-40357, Ataxin-2 shRNA Plasmid (h): sc-40356-SH, Ataxin-2 shRNA Plasmid (m): sc-40357-SH, Ataxin-2 shRNA (h) Lentiviral Particles: sc-40356-V and Ataxin-2 shRNA (m) Lentiviral Particles: sc-40357-V.

Molecular Weight of (predicted) Ataxin-2: 140 kDa.

Molecular Weight of (observed) Ataxin-2: 160 kDa.

DATA



Ataxin-2 (C-18): sc-18478. Immunoperoxidase staining of formalin fixed, paraffin-embedded human cerebral cortex tissue showing cytoplasmic staining of neuronal cells and glial cells.

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 or our catalog for detailed protocols and support products.


 MONOS
 Satisfaction
 Guaranteed

Try **Ataxin-2 (A-6): sc-515602**, our highly recommended monoclonal alternative to Ataxin-2 (C-18).