

Ataxin-2 (A-6): sc-515602

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/ATXN2) 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

1. Gispert, S., et al. 1993. Chromosomal assignment of the second locus for autosomal dominant cerebellar ataxia (SCA2) to chromosome 12q23-24.1 *Nat. Genet.* 4: 295-299.
2. Sanpei, K., et al. 1996. Identification of the spinocerebellar ataxia type 2 gene using a direct identification of repeat expansion and cloning technique, DIRECT. *Nat. Genet.* 14: 277-284.
3. Pujana, M.A., et al. 1999. Spinocerebellar ataxias in Spanish patients: genetic analysis of familial and sporadic cases. *The Ataxia Study Group. Hum. Genet.* 104: 516-522.
4. 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.

CHROMOSOMAL LOCATION

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

SOURCE

Ataxin-2 (A-6) is a mouse monoclonal antibody specific for an epitope mapping between amino acids 1086-1113 within an internal region of Ataxin-2 of human origin.

PRODUCT

Each vial contains 200 µg IgM kappa light chain in 1.0 ml of PBS with < 0.1% sodium azide and 0.1% gelatin.

Blocking peptide available for competition studies, sc-515602 P, (100 µg peptide in 0.5 ml PBS containing < 0.1% sodium azide and 0.2% stabilizer protein).

STORAGE

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

APPLICATIONS

Ataxin-2 (A-6) is recommended for detection of Ataxin-2 of mouse, rat and human origin by Western Blotting (starting dilution 1:100, 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).

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 (predicted) of Ataxin-2: 140 kDa.

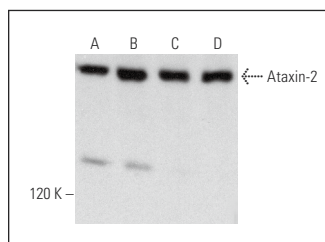
Molecular Weight (observed) of Ataxin-2: 160 kDa.

Positive Controls: Hep G2 cell lysate: sc-2227, DU 145 cell lysate: sc-2268 or SK-N-MC cell lysate: sc-2237.

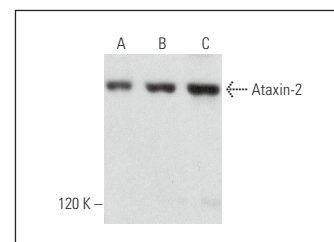
RECOMMENDED SUPPORT REAGENTS

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) Immunoprecipitation: use Protein L-Agarose: sc-2336 (0.5 ml agarose/2.0 ml). 3) 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.

DATA



Ataxin-2 (A-6): sc-515602. Western blot analysis of Ataxin-2 expression in Jurkat (A), K-562 (B), HeLa (C) and PANC-1 (D) whole cell lysates.



Ataxin-2 (A-6): sc-515602. Western blot analysis of Ataxin-2 expression in Hep G2 (A), DU 145 (B) and SK-N-MC (C) whole cell lysates.

SELECT PRODUCT CITATIONS

1. Cascella, R., et al. 2022. A quantitative biology approach correlates neuronal toxicity with the largest inclusions of TDP-43. *Sci. Adv.* 8: eabm6376.

RESEARCH USE

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