SANTA CRUZ BIOTECHNOLOGY, INC.

ATXN7L2 (G-14): sc-137306

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The Power to Question

BACKGROUND

SCA7 is an autosomal dominant neurodegenerative disorder characterized by ataxia and selective neuronal cell loss caused by the expansion of a translated CAG repeat encoding a polyglutamine tract in ataxin-7, which is the SCA7 gene product. Ataxin-7 is a nuclear protein that is expressed within neurons both affected and unaffected in SCA7 pathology with subcellular localization being variable depending upon the neuronal subtype. Polyglutamine expanded in ataxin-7 may carry out its pathogenic effects in the nucleus by altering the matrix-associated nuclear structure and/or by disrupting nucleolar function.ATXN7L2 (Ataxin-7-like protein 2) is a 722 amino acid protein that contains a SCA7 domain, which is highly conserved through all members of the ATXN7 gene family. The gene encoding ATXN7L2 maps to human chromosome 1, the largest human chromosome spanning about 260 million base pairs and making up 8% of the human genome.

REFERENCES

1. Holmberg, M., et al. 1998. Spinocerebellar ataxia type 7 (SCA7): a neurodegenerative disorder with neuronal intranuclear inclusions. Hum. Mol. Genet. 7: 913-918.
2. Chen, S., et al. 2004. Interference of Crx-dependent transcription by ataxin-7 involves interaction between the glutamine regions and requires the ataxin-7 carboxy-terminal region for nuclear localization. Hum. Mol. Genet. 13: 53-67.
3. Gregory, S.G., et al. 2006. The DNA sequence and biological annotation of human chromosome 1. Nature 441: 315-321.
4. Garden, G.A. and La Spada, A.R. 2008. Molecular pathogenesis and cellular pathology of spinocerebellar ataxia type 7 neurodegeneration. Cerebellum 7: 138-149.
5. Zhao, Y., et al. 2008. A TFTC/STAGA module mediates histone H2A and H2B deubiquitination, coactivates nuclear receptors, and counteracts heterochromatin silencing. Mol. Cell. 29: 92-101.
6. Lin, Y., et al. 2008. Trinucleotide expansions in the SCA7 gene in a large family with spinocerebellar ataxia and craniocervical dystonia. Neurosci. Lett. 434: 230-233.
7. Hugosson, T., et al. 2009. Macular dysfunction and morphology in spinocerebellar ataxia type 7 (SCA 7). Ophthalmic Genet. 30: 1-6.
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CHROMOSOMAL LOCATION

Genetic locus: ATXN7L2 (human) mapping to 1p13.3; Atxn7l2 (mouse) mapping to 3 F2.3.

SOURCE

ATXN7L2 (G-14) is an affinity purified goat polyclonal antibody raised against a peptide mapping within an internal region of ATXN7L2 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-137306 P, (100 µg peptide in 0.5 ml PBS containing < 0.1% sodium azide and 0.2% BSA).

APPLICATIONS

ATXN7L2 (G-14) is recommended for detection of ATXN7L2 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); non cross-reactive with other ATXN family members.

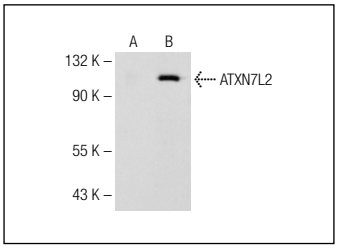
Suitable for use as control antibody for ATXN7L2 siRNA (h): sc-78857, ATXN7L2 siRNA (m): sc-141379, ATXN7L2 shRNA Plasmid (h): sc-78857-SH, ATXN7L2 shRNA Plasmid (m): sc-141379-SH, ATXN7L2 shRNA (h) Lentiviral Particles: sc-78857-V and ATXN7L2 shRNA (m) Lentiviral Particles: sc-141379-V.

Molecular Weight (predicted) of ATXN7L2: 77 kDa.

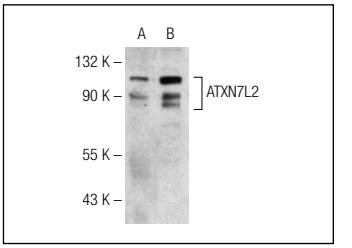
Molecular Weight (observed) of ATXN7L2: 110 kDa.

Positive Controls: ATXN7L2 (h): 293T Lysate: sc-115673.

DATA



ATXN7L2 (G-14): sc-137306. Western blot analysis of ATXN7L2 expression in non-transfected: sc-117752 (**A**) and human ATXN7L2 transfected: sc-115673 (**B**) 293T whole cell lysates.



ATXN7L2 (G-14): sc-137306. Western blot analysis of ATXN7L2 expression in mouse liver tissue extract (**A**) and LADMAC whole cell lysate (**B**).

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.