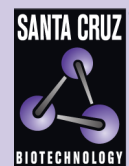


SETX (QQ-7): sc-100319



The Power to Question

BACKGROUND

SETX (senataxin), also known as ALS4 (amyotrophic lateral sclerosis 4 protein), AOA2 or SCAR1, belongs to the DNA2/NAM7 helicase family. Localizing to the nucleolus or the nucleoplasm in a cell cycle-dependent manner and to the cytoplasm, SETX contains a C-terminal DNA/RNA helicase domain and is believed to function as a helicase involved in RNA processing and DNA repair. Mutations in the gene encoding SETX can lead to ataxia-ocular apraxia 2 (AOA2) or amyotrophic lateral sclerosis 4 (ALS4). AOA2, also known as spinocerebellar ataxia-1 (SCAR1), is an autosomal recessive disorder characterized by progressive neurodegeneration of the cerebellum associated with the loss of Purkinje cells. ALS4 is a familial childhood- or adolescent-onset neurodegenerative disorder affecting both upper and lower motor neurons that ultimately results in fatal paralysis.

REFERENCES

1. Online Mendelian Inheritance in Man, OMIM™. 2002. Johns Hopkins University, Baltimore, MD. MIM Number: 602433. World Wide Web URL: <http://www.ncbi.nlm.nih.gov/omim/>
2. Moreira, M.C., et al. 2004. Senataxin, the ortholog of a yeast RNA helicase, is mutant in ataxia-ocular apraxia 2. *Nat. Genet.* 36: 225-227.
3. Chen, Y.Z., et al. 2006. Senataxin, the yeast Sen1p orthologue: characterization of a unique protein in which recessive mutations cause ataxia and dominant mutations cause motor neuron disease. *Neurobiol. Dis.* 23: 97-108.
4. Asaka, T., et al. 2006. Autosomal recessive ataxia with peripheral neuropathy and elevated AFP: novel mutations in SETX. *Neurology* 66: 1580-1581.
5. Nahas, S.A., et al. 2007. Ataxia-oculomotor apraxia 2 patients show no increased sensitivity to ionizing radiation. *Neuromuscul. Disord.* 17: 968-969.
6. Suraweera, A., et al. 2007. Senataxin, defective in ataxia oculomotor apraxia type 2, is involved in the defense against oxidative DNA damage. *J. Cell Biol.* 177: 969-979.

CHROMOSOMAL LOCATION

Genetic locus: SETX (human) mapping to 9q34.13; Setx (mouse) mapping to 2 A3.

SOURCE

SETX (QQ-7) is a mouse monoclonal antibody raised against a partial recombinant protein mapping within amino acids 2579-2676 of SETX of human origin.

PRODUCT

Each vial contains 100 µg IgG₁ kappa light chain 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.

APPLICATIONS

SETX (QQ-7) is recommended for detection of SETX 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).

Suitable for use as control antibody for SETX siRNA (h): sc-92842, SETX siRNA (m): sc-153389, SETX shRNA Plasmid (h): sc-92842-SH, SETX shRNA Plasmid (m): sc-153389-SH, SETX shRNA (h) Lentiviral Particles: sc-92842-V and SETX shRNA (m) Lentiviral Particles: sc-153389-V.

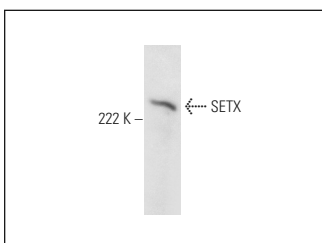
Molecular Weight of SETX: 303 kDa.

Positive Controls: NIH/3T3 whole cell lysate: sc-2210.

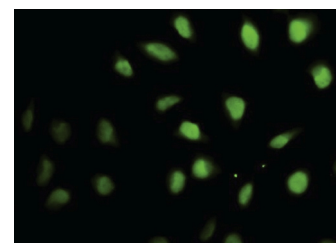
RECOMMENDED SUPPORT REAGENTS

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 A/G PLUS-Agarose: sc-2003 (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



SETX (QQ-7): sc-100319. Western blot analysis of SETX expression in NIH/3T3 whole cell lysate.



SETX (QQ-7): sc-100319. Immunofluorescence staining of paraformaldehyde-fixed HeLa cells showing nuclear localization.

SELECT PRODUCT CITATIONS

1. Vantaggiato, C., et al. 2011. Senataxin modulates neurite growth through fibroblast growth factor 8 signalling. *Brain* 134: 1808-1828.
2. Lei, L., et al. 2021. Unusual electrophysiological findings in a Chinese ALS 4 family with SETX-L389S mutation: a three-year follow-up. *J. Neurol.* 268: 1050-1058.

RESEARCH USE

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