

Spastin (D-20): sc-49525

BACKGROUND

The AAA protein family members share an ATPase domain and have roles in various cellular processes including intracellular motility, membrane trafficking, proteolysis, protein folding and organelle biogenesis. Spastin, a member of the AAA protein family, is a 616 amino acid protein and is involved in the function or assembly of nuclear protein complexes. The Spastin protein is expressed ubiquitously and localizes to the nucleus and the cytoplasm, where it may also be involved in microtubule dynamics. Mutations in the Spastin gene (SPAST, SPG4) cause the most common form of spastic paraplegia 4, an autosomal dominant form of hereditary spastic paraplegia (HSP). HSPs comprise a group of inherited neurological disorders characterized by spastic lower extremity weakness due to a length-dependent, retrograde axonopathy of corticospinal motor neurons. SPAST-specific mutations account for approximately 40% of all autosomal dominant HSPs.

REFERENCES

- Nielsen, J.E., et al. 2004. Hereditary spastic paraplegia with cerebellar ataxia: a complex phenotype associated with a new SPG4 gene mutation. *Eur. J. Neurol.* 11: 8178-8124.
- Scheuer, K.H., et al. 2005. Reduced regional cerebral blood flow in SPG4-linked hereditary spastic paraplegia. *J. Neurol. Sci.* 235: 23-32.
- Alber, B., et al. 2005. Spastin related hereditary spastic paraplegia with dysplastic corpus callosum. *J. Neurol. Sci.* 236: 9-12.
- Claudiani, P., et al. 2005. Spastin subcellular localization is regulated through usage of different translation start sites and active export from the nucleus. *Exp. Cell Res.* 309: 358-369.
- Meyer, T., et al. 2005. Early-onset ALS with long-term survival associated with spastin gene mutation. *Neurology* 65: 141-143.
- Svenson, I.K., et al. 2005. Subcellular localization of Spastin: implications for the pathogenesis of hereditary spastic paraplegia. *Neurogenetics* 6: 135-141.
- Winner, B., et al. 2006. Thin corpus callosum and amyotrophy in spastic paraplegia—Case report and review of literature. *Clin. Neurol. Neurosurg.* 108: 692-698.
- Depienne, C., et al. 2006. Spastin mutations are frequent in sporadic spastic paraparesis and their spectrum is different from the one observed in familial cases. *J. Med. Genet.* 43: 259-265.

CHROMOSOMAL LOCATION

Genetic locus: SPAST (human) mapping to 2p22.3; Spg4 (mouse) mapping to 17 E2.

SOURCE

Spastin (D-20) is an affinity purified goat polyclonal antibody raised against a peptide mapping within an internal region of Spastin of human origin.

STORAGE

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

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-49525 P, (100 µg peptide in 0.5 ml PBS containing < 0.1% sodium azide and 0.2% BSA).

APPLICATIONS

Spastin (D-20) is recommended for detection of Spastin 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) and solid phase ELISA (starting dilution 1:30, dilution range 1:30-1:3000); not recommended for the detection of isoform 2.

Spastin (D-20) is also recommended for detection of Spastin in additional species, including equine, canine, bovine, porcine and avian.

Suitable for use as control antibody for Spastin siRNA (h): sc-61603, Spastin siRNA (m): sc-61604, Spastin shRNA Plasmid (h): sc-61603-SH, Spastin shRNA Plasmid (m): sc-61604-SH, Spastin shRNA (h) Lentiviral Particles: sc-61603-V and Spastin shRNA (m) Lentiviral Particles: sc-61604-V.

Molecular Weight of Spastin long isoform: 64-68 kDa.

Molecular Weight of Spastin short isoform: 55-60 kDa.

Positive Controls: HeLa whole cell lysate: sc-2200.

RECOMMENDED SECONDARY REAGENTS

To ensure optimal results, the following support (secondary) reagents are recommended: 1) Western Blotting: use donkey anti-goat IgG-HRP: sc-2020 (dilution range: 1:2000-1:100,000) or Cruz Marker™ compatible donkey anti-goat IgG-HRP: sc-2033 (dilution range: 1:2000-1:5000), Cruz Marker™ Molecular Weight Standards: sc-2035, TBS Blotto A Blocking Reagent: sc-2333 and Western Blotting Luminol Reagent: sc-2048. 2) Immunofluorescence: use donkey anti-goat IgG-FITC: sc-2024 (dilution range: 1:100-1:400) or donkey anti-goat IgG-TR: sc-2783 (dilution range: 1:100-1:400) with UltraCruz™ Mounting Medium: sc-24941.

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.



Try **Spastin (C-3): sc-374068** or **Spastin (Sp 3G11/1): sc-53443**, our highly recommended monoclonal alternatives to Spastin (D-20).