

# Spastin (A-2): sc-271247

## 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.

## SOURCE

Spastin (A-2) is a mouse monoclonal antibody raised against amino acids 61-194 mapping near the N-terminus of Spastin of human origin.

## PRODUCT

Each vial contains 200 µg IgG<sub>2a</sub> kappa light chain in 1.0 ml of PBS with < 0.1% sodium azide and 0.1% gelatin.

## APPLICATIONS

Spastin (A-2) is recommended for detection of Spastin of 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 Spastin siRNA (h): sc-61603, Spastin shRNA Plasmid (h): sc-61603-SH and Spastin shRNA (h) Lentiviral Particles: sc-61603-V.

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

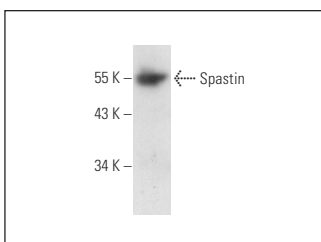
Molecular Weight of Spastin short isoform: 55-60 kDa.

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

## RECOMMENDED 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



Spastin (A-2): sc-271247. Western blot analysis of Spastin expression in HeLa whole cell lysate.

## SELECT PRODUCT CITATIONS

- Zhu, Z., et al. 2019. Novel mutations in the SPAST gene cause hereditary spastic paraplegia. *Parkinsonism Relat. Disord.* 69: 125-133.

## 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.