SANTA CRUZ BIOTECHNOLOGY, INC.

Paraplegin (S-12): sc-55978

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The Power to Question

BACKGROUND

Paraplegin, also known as SPG7 (spastic paraplegia protein 7), CAR, CMAR or PGN, is a 795 amino acid metalloprotease that is a member of the AAA protein family. Localized to the mitochrondrial membrane and expressed throughout the body, Paraplegin is a multi-pass membrane protein that is thought to be involved in signal transduction and chaperone-like activities in the mitochrondria. Defects in the gene encoding Paraplegin are the cause of spastic paraplegia type 7 (SPG7), a form of autosomal recessive hereditary spastic paraplegia (AR-HSP). HSPs are degenerative spinal cord disorders that are characterized by muscle spasms, stiffness in the legs and, in some cases, incontinence. Recent studies suggest that SPG7 may be a mitochondrial-based disease, as mutations in the Paraplegin gene lead to ragged-red fibers, oxidase-negative fibers and intense succinate dehydrogenase-stained areas of the mitochrondria. These mitochondrial dysfunctions lead to axonal degeneration and impaired axonal transport, thus causing the neurodegeneration seen in HSPs.

REFERENCES

1. Settasatian, C., et al. 1999. Genomic structure and expression analysis of the spastic paraplegia gene, SPG7. Hum. Genet. 105: 139-144.
2. Wilkinson, P.A., et al. 2004. A clinical, genetic and biochemical study of SPG7 mutations in hereditary spastic paraplegia. Brain 127: 973-980.
3. Lindholm, D., et al. 2004. Mitochondrial proteins in neuronal degeneration. Biochem. Biophys. Res. Commun. 321: 753-758.
4. Claypool, S.M. and Koehler, C.M. 2005. Hereditary spastic paraplegia: respiratory choke or unactivated substrate? Cell 123: 183-185.
5. Nolden, M., et al. 2005. The m-AAA protease defective in hereditary spastic paraplegia controls ribosome assembly in mitochondria. Cell 123: 277-289.
6. Pirozzi, M., et al. 2006. Intramuscular viral delivery of paraplegin rescues peripheral axonopathy in a model of hereditary spastic paraplegia. J. Clin. Invest. 116: 202-208.
7. Elleuch, N., et al. 2006. Mutation analysis of the paraplegin gene (SPG7) in patients with hereditary spastic paraplegia. Neurology 66: 654-659.
8. Warnecke, T., et al. 2007. A novel form of autosomal recessive hereditary spastic paraplegia caused by a new SPG7 mutation. Neurology 69: 368-375.
9. Arnoldi, A., et al. 2008. A clinical, genetic, and biochemical characterization of SPG7 mutations in a large cohort of patients with hereditary spastic paraplegia. Hum. Mutat. 29: 522-531.

CHROMOSOMAL LOCATION

Genetic locus: SPG7 (human) mapping to 16q24.3; Spg7 (mouse) mapping to 8 E1.

SOURCE

Paraplegin (S-12) is an affinity purified goat polyclonal antibody raised against a peptide mapping within an internal region of Paraplegin 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-55978 P, (100 µg peptide in 0.5 ml PBS containing < 0.1% sodium azide and 0.2% BSA).

APPLICATIONS

Paraplegin (S-12) is recommended for detection of Paraplegin 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).

Paraplegin (S-12) is also recommended for detection of Paraplegin in additional species, including equine, canine and bovine.

Suitable for use as control antibody for Paraplegin siRNA (h): sc-62755, Paraplegin siRNA (m): sc-62756, Paraplegin shRNA Plasmid (h): sc-62755-SH, Paraplegin shRNA Plasmid (m): sc-62756-SH, Paraplegin shRNA (h) Lentiviral Particles: sc-62755-V and Paraplegin shRNA (m) Lentiviral Particles: sc-62756-V.

Molecular Weight of Paraplegin: 88 kDa.

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.

SELECT PRODUCT CITATIONS

1. Bahat, A., et al. 2014. StAR enhances transcription of genes encoding the mitochondrial proteases involved in its own degradation. Mol. Endocrinol. 28: 208-224.

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.