SANTA CRUZ BIOTECHNOLOGY, INC.

Paraplegin (C-5): sc-514393

SANTA CRUZ BIOTECHNOLOGY

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.

CHROMOSOMAL LOCATION

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

SOURCE

Paraplegin (C-5) is a mouse monoclonal antibody raised against amino acids 131-310 mapping within an internal region of Paraplegin of human origin.

PRODUCT

Each vial contains 200 µg IgG_{2b} kappa light chain in 1.0 ml of PBS with < 0.1% sodium azide and 0.1% gelatin.

Paraplegin (C-5) is available conjugated to agarose (sc-514393 AC), 500 µg/ 0.25 ml agarose in 1 ml, for IP; to HRP (sc-514393 HRP), 200 µg/ml, for WB, IHC(P) and ELISA; to either phycoerythrin (sc-514393 PE), fluorescein (sc-514393 FITC), Alexa Fluor® 488 (sc-514393 AF488), Alexa Fluor® 546 (sc-514393 AF546), Alexa Fluor® 594 (sc-514393 AF594) or Alexa Fluor® 647 (sc-514393 AF647), 200 µg/ml, for WB (RGB), IF, IHC(P) and FCM; and to either Alexa Fluor® 680 (sc-514393 AF680) or Alexa Fluor® 790 (sc-514393 AF790), 200 µg/ml, for Near-Infrared (NIR) WB, IF and FCM.

STORAGE

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

APPLICATIONS

Paraplegin (C-5) is recommended for detection of Paraplegin of mouse, rat and 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).

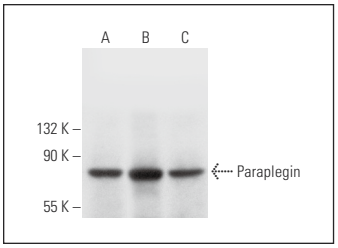
Suitable for use as control antibody for Paraplegin siRNA (h): sc-62755, Paraplegin siRNA (m): sc-62756, Paraplegin shRNA Plasmid (h): sc-62755-SH, Paraplegin shRNA Plasmid (m): sc-62756-SH, Paraplegin shRNA (h) Lentiviral Particles: sc-62755-V and Paraplegin shRNA (m) Lentiviral Particles: sc-62756-V.

Molecular Weight of Paraplegin isoform 1: 88 kDa.

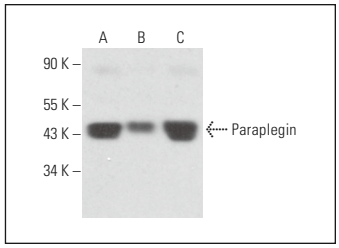
Molecular Weight of Paraplegin isoform 2: 54 kDa.

Positive Controls: HeLa whole cell lysate: sc-2200, Hep G2 cell lysate: sc-2227 or JAR cell lysate: sc-2276.

DATA



Paraplegin (C-5): sc-514393. Western blot analysis of Paraplegin expression in HeLa (**A**), Hep G2 (**B**) and JAR (**C**) whole cell lysates.



Paraplegin (C-5): sc-514393. Western blot analysis of Paraplegin expression in Hep G2 (**A**), MIA PaCa-2 (**B**) and A549 (**C**) whole cell lysates.

SELECT PRODUCT CITATIONS

1. Tsai, C.W., et al. 2017. Proteolytic control of the mitochondrial calcium uniporter complex. Proc. Natl. Acad. Sci. USA 114: 4388-4393.
2. Verdura, E., et al. 2020. A deep intronic splice variant advises reexamination of presumably dominant SPG7 cases. Ann. Clin. Transl. Neurol. 7: 105-111.
3. Aishwarya, R., et al. 2020. Pleiotropic effects of mdivi-1 in altering mitochondrial dynamics, respiration, and autophagy in cardiomyocytes. Redox Biol. 36: 101660.
4. Liu, H., et al. 2022. Prohibitin 1 regulates mtDNA release and downstream inflammatory responses. EMBO J. 41: e111173.
5. Aishwarya, R., et al. 2024. Diastolic dysfunction in Alzheimer's disease model mice is associated with Aβ-Amyloid aggregate formation and mitochondrial dysfunction. Sci. Rep. 14: 16715.

RESEARCH USE

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