

Paraplegin (G-16): sc-55976

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 mitochondrial 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 mitochondria. 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 mitochondria. These mitochondrial dysfunctions lead to axonal degeneration and impaired axonal transport, thus causing the neurodegeneration seen in HSPs.

REFERENCES

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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.
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CHROMOSOMAL LOCATION

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

SOURCE

Paraplegin (G-16) 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-55976 P, (100 µg peptide in 0.5 ml PBS containing < 0.1% sodium azide and 0.2% BSA).

APPLICATIONS

Paraplegin (G-16) 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 (G-16) 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 isoform 1: 88 kDa.

Molecular Weight of Paraplegin isoform 2: 54 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.

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.



Try **Paraplegin (C-5): sc-514393**, our highly recommended monoclonal alternative to Paraplegin (G-16).