

mucolipin 1 (L-15): sc-26266

BACKGROUND

The gene encoding human mucolipin 1 maps to chromosome 19p13.2. Mutations in this gene cause a rare, autosomal recessive lysosomal storage disease known as mucopolipidosis type IV (MLIV). Clinical characteristics of MLIV include psychomotor retardation, retinal degeneration, corneal opacities and strabismus. Mucolipin 1 localizes to the plasma membrane and contains six transmembrane domains. The carboxy-terminus of mucolipin 1 shares sequence homology with polycystin-2 and the transient receptor potential cation channel family. The concentration of intracellular Ca²⁺ regulates the permeability of mucolipin 1 to Ca²⁺, Na⁺ and K⁺. The influence of Ca²⁺ on mucolipin 1 represents a possible role for mucolipin 1 in lysosomal exocytosis and the trafficking of late endosomes and lysosomes.

REFERENCES

- Merin, S., et al. 1975. Mucopolipidosis IV: ocular, systemic, and ultrastructural findings. *Invest. Ophthalmol.* 14: 437-448.
- Bargal, R., et al. 2000. Identification of the gene causing mucopolipidosis type IV. *Nat. Genet.* 26: 118-123.
- Sun, M., et al. 2000. Mucopolipidosis type IV is caused by mutations in a gene encoding a novel transient receptor potential channel. *Hum. Mol. Genet.* 9: 2471-2478.

SOURCE

mucolipin 1 (L-15) is an affinity purified goat polyclonal antibody raised against a peptide mapping within an internal region of mucolipin 1 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-26266 P, (100 µg peptide in 0.5 ml PBS containing < 0.1% sodium azide and 0.2% BSA).

APPLICATIONS

mucolipin 1 (L-15) is recommended for detection of mucolipin 1 and, to a lesser extent, mucolipin 2 and 3 of mouse, rat and human origin by Western Blotting (starting dilution 1:200, 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).

mucolipin 1 (L-15) is also recommended for detection of mucolipin 1 and, to a lesser extent, mucolipin 2 and 3 in additional species, including equine, canine, bovine and avian.

Suitable for use as control antibody for mucolipin 1 siRNA (h): sc-44519 and mucolipin 1 siRNA (m): sc-44520.

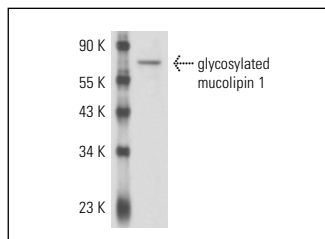
Molecular Weight of NRSF: 65 kDa.

Positive Controls: Jurkat whole cell lysate: sc-2204.

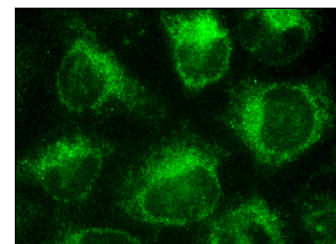
RECOMMENDED SECONDARY REAGENTS

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) Immunoprecipitation: use Protein A/G PLUS-Agarose: sc-2003 (0.5 ml agarose/2.0 ml). 3) 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.

DATA



mucolipin 1 (L-15): sc-26266. Western blot analysis of mucolipin 1 expression in Jurkat whole cell lysate.



mucolipin 1 (L-15): sc-26266. Immunofluorescence staining of methanol-fixed HeLa cells showing cytoplasmic localization.

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.



MONOS
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Guaranteed

Try **mucolipin 1 (F-10): sc-398868**, our highly recommended monoclonal alternative to mucolipin 1 (L-15).