Product Information

3050 Spruce Street, Saint Louis, MO 63103 USA
Tel: (800) 521-8956 (314) 771-5765 Fax: (800) 325-5052 (314) 771-5757
email: techservice@sial.com sigma-aldrich.com

Anti-Mucolipin-3 (C-terminal)
produced in rabbit, affinity isolated antibody

Catalog Number **M1071**

Product Description
Anti-Mucolipin-3 (C-terminal) is produced in rabbit using as immunogen a synthetic peptide corresponding to amino acids 529-542 of human mucolipin-3 (GeneID: 55283), conjugated to KLH. This sequence is identical in mouse and rat mucolipin-3. The antibody is affinity-purified using the immunizing peptide immobilized on agarose.

Anti-Mucolipin-3 (C-terminal) specifically recognizes human mucolipin-3 by immunoblotting (~75 kDa). Additional bands may be observed at ~150 kDa and ~50 kDa, due to aggregation and degradation of mucolipin-3, respectively. Staining of the mucolipin-3 band is specifically inhibited by the immunizing peptide.

Mucolipidosis type IV (MLIV) is an autosomal recessive, neurodegenerative disorder caused by mutations in the MCOLN1 gene that encodes mucolipin-1 (also termed TRP-ML1, MLN1, ML1 mucolipidin).[1, 2] MLVI is associated with severe psychomotor retardation and ophthalmologic defects. MLIV is a lysosomal storage disorder associated with lysosomal accumulation of sphingolipids, phospholipids and mucopolysaccharides. Unlike other mucolipidoses, lysosomal hydrolase activity is not impaired in MLVI. Rather, MLVI pathophysiology has been linked to deficiency in membrane trafficking and organelle dynamics in the late endocytic pathway.[2] MLN1 shares significant sequence homology with the TRP superfamily of cation channels, characterized by permeability to monovalent cations and Ca^{2+}. MLN1 is thought to function as a proton-leak channel in lysosomes, regulating lysosomal pH and hydrolytic activity.[3] MLN1 has been localized to late endosomes and lysosomes. In addition to MLN1, mammals encode two other highly related proteins, MLN2/TRPML2 and MLN3/TRPML3. Mutations in mouse mucolipin-3 (MLN3, TRPML3) encoded by the MCOLN3 gene, are associated with deafness and pigmentation defects in varitint-waddler mice.[4] TRPMLs have been shown interact to form homo- and heteromultimers resulting in a complex pattern of localization in both the lysosomal and endoplasmic reticulum compartments.[5]

Reagent
Supplied as a solution in 0.01 M phosphate buffered saline, pH 7.4, containing 15 mM sodium azide as a preservative.

Antibody concentration: ~1.5 mg/mL

Precautions and Disclaimer
This product is for R&D use only, not for drug, household, or other uses. Please consult the Material Safety Data Sheet for information regarding hazards and safe handling practices.

Storage/Stability
For continuous use, store at 2-8 °C for up to one month. For extended storage, freeze in working aliquots. Repeated freezing and thawing, or storage in "frost-free" freezers, is not recommended. If slight turbidity occurs upon prolonged storage, clarify the solution by centrifugation before use. Working dilution samples should be discarded if not used within 12 hours.

Product Profile
Immunoblotting: a working antibody concentration of 0.5-1.0 μg/mL is recommended using HEK-293T cells expressing human mucolipin-3.

Note: In order to obtain the best results and assay sensitivity in various techniques and preparations, we recommend determining optimal working concentrations by titration.

References
1. Bach, G., et al., *Eur. J. Physiol.*, **451**, 313-317 (2005).
2. Bargal, R., et al., *Nature Genet.*, **26**, 118-123 (2000).
3. Soyombo, A.A., et al., *J. Biol. Chem.,* **281**, 7294-7301 (2006).
4. Di Palma, F., et al., *Proc. Natl. Acad. Sci. USA*, **99**, 14994-14999 (2002).
5. Zeevi, D.A., et al., *Biochim. Biophys. Acta,* **1772**, 851-858 (2007).

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