

SUMF1 (A-4): sc-374102

BACKGROUND

Sulfatases are enzymes that remove sulfate residues from a variety of substrates via the hydrolysis of sulfate esters. In order to function properly, sulfatases require the presence of C α -formylglycine (FGly), a unique amino acid, in their active site. This amino acid is synthesized by enzymes that use a cysteine to posttranslationally generate FGly. SUMF1 (sulfatase modifying factor 1), also known as FGE, is a 374 amino acid alternatively spliced protein that localizes to the lumen of the endoplasmic reticulum and belongs to the sulfatase-modifying factor family. Expressed ubiquitously with highest expression in liver, kidney and pancreas, SUMF1 exists as either a monomer, a homodimer or a heterodimer (with SUMF2) and functions to oxidize sulfatase cysteine residues to an active FGly residue, thereby playing an important role in sulfatase activity. Defects in the gene encoding SUMF1 are the cause of multiple sulfatase deficiency (MSD), a heterogeneous disorder characterized by metachromatic leukodystrophy, mucopolysaccharidosis, chondrodysplasia punctata, hydrocephalus, ichthyosis, neurologic deterioration and developmental delay.

REFERENCES

1. Cosma, M.P., et al. 2003. The multiple sulfatase deficiency gene encodes an essential and limiting factor for the activity of sulfatases. *Cell* 113: 445-456.
2. Zito, E., et al. 2005. Sulphatase activities are regulated by the interaction of sulphatase-modifying factor 1 with SUMF2. *EMBO Rep.* 6: 655-660.
3. Fraldi, A., et al. 2007. SUMF1 enhances sulfatase activities *in vivo* in five sulfatase deficiencies. *Biochem. J.* 403: 305-312.
4. Zito, E., et al. 2007. Sulfatase modifying factor 1 trafficking through the cells: from endoplasmic reticulum to the endoplasmic reticulum. *EMBO J.* 26: 2443-2453.
5. Annunziata, I., et al. 2007. Multiple sulfatase deficiency is due to hypomorphic mutations of the SUMF1 gene. *Hum. Mutat.* 28: 928.
6. Online Mendelian Inheritance in Man, OMIM™. 2007. Johns Hopkins University, Baltimore, MD. MIM Number: 607939. World Wide Web URL: <http://www.ncbi.nlm.nih.gov/omim/>
7. Fraldi, A., et al. 2008. Multistep, sequential control of the trafficking and function of the multiple sulfatase deficiency gene product, SUMF1 by pDI, ERGIC-53 and ERp44. *Hum. Mol. Genet.* 17: 2610-2621.
8. Schlotawa, L., et al. 2008. Molecular analysis of SUMF1 mutations: stability and residual activity of mutant formylglycine-generating enzyme determine disease severity in multiple sulfatase deficiency. *Hum. Mutat.* 29: 205.

CHROMOSOMAL LOCATION

Genetic locus: SUMF1 (human) mapping to 3p26.1; Sumf1 (mouse) mapping to 6 E1.

SOURCE

SUMF1 (A-4) is a mouse monoclonal antibody raised against amino acids 225-355 mapping near the C-terminus of SUMF1 of human origin.

PRODUCT

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

APPLICATIONS

SUMF1 (A-4) is recommended for detection of SUMF1 of mouse, rat and human origin by Western Blotting (starting dilution 1:100, dilution range 1:100-1:1000), immunoprecipitation [1-2 μ g per 100-500 μ g 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 SUMF1 siRNA (h): sc-76610, SUMF1 siRNA (m): sc-76611, SUMF1 shRNA Plasmid (h): sc-76610-SH, SUMF1 shRNA Plasmid (m): sc-76611-SH, SUMF1 shRNA (h) Lentiviral Particles: sc-76610-V and SUMF1 shRNA (m) Lentiviral Particles: sc-76611-V.

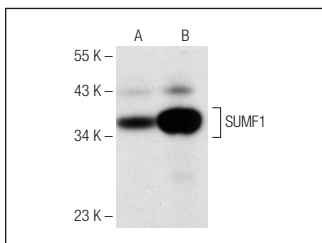
Molecular Weight of SUMF1: 42 kDa.

Positive Controls: Caki-1 cell lysate: sc-2224 or Hep G2 cell lysate: sc-2227.

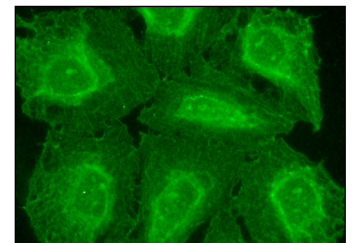
RECOMMENDED SUPPORT REAGENTS

To ensure optimal results, the following support reagents are recommended: 1) Western Blotting: use m-IgG κ BP-HRP: sc-516102 or m-IgG κ BP-HRP (Cruz Marker): sc-516102-CM (dilution range: 1:1000-1:10000), Cruz Marker™ Molecular Weight Standards: sc-2035, UltraCruz® Blocking Reagent: sc-516214 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 m-IgG κ BP-FITC: sc-516140 or m-IgG κ BP-PE: sc-516141 (dilution range: 1:50-1:200) with UltraCruz® Mounting Medium: sc-24941 or UltraCruz® Hard-set Mounting Medium: sc-359850.

DATA



SUMF1 (A-4): sc-374102. Western blot analysis of SUMF1 expression in Caki-1 (A) and Hep G2 (B) whole cell lysates.



SUMF1 (A-4): sc-374102. Immunofluorescence staining of methanol-fixed HeLa cells showing membrane and cytoskeletal 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.