



Rabbit Anti-SUMF1 antibody

SL12366R

Product Name:	SUMF1
Chinese Name:	硫酸酯酶修饰因子1抗体
Alias:	MGC150436; AAPA3037; C alpha formylglycine generating enzyme 1; C-alpha-formylglycine-generating enzyme 1; FGE; FGly generating enzyme; MGC131853; Sulfatase modifying factor 1 [Precursor]; Sulfatase-modifying factor 1; SUMF1; SUMF1 HUMAN; UNQ3037.
Organism Species:	Rabbit
Clonality:	Polyclonal
React Species:	Human,Mouse,Rat,Dog,Pig,Cow,Horse,Rabbit,Sheep,
Applications:	WB=1:500-2000ELISA=1:500-1000IHC-P=1:400-800IHC-F=1:400-800ICC=1:100-500IF=1:100-500 (Paraffin sections need antigen repair) not yet tested in other applications. optimal dilutions/concentrations should be determined by the end user.
Molecular weight:	37kDa
Cellular localization:	cytoplasmic
Form:	Lyophilized or Liquid
Concentration:	1mg/ml
immunogen:	KLH conjugated synthetic peptide derived from human SUMF1:301-374/374
Lsotype:	IgG
Purification:	affinity purified by Protein A
Storage Buffer:	0.01M TBS(pH7.4) with 1% BSA, 0.03% Proclin300 and 50% Glycerol.
Storage:	Store at -20 °C for one year. Avoid repeated freeze/thaw cycles. The lyophilized antibody is stable at room temperature for at least one month and for greater than a year when kept at -20°C. When reconstituted in sterile pH 7.4 0.01M PBS or diluent of antibody the antibody is stable for at least two weeks at 2-4 °C.
PubMed:	PubMed
Product Detail:	SUMF1 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 FGIy 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.

Function:

Using molecular oxygen and an unidentified reducing agent, oxidizes a cysteine residue in the substrate sulfatase to an active site 3-oxoalanine residue, which is also called C(alpha)-formylglycine. Known substrates include GALNS, ARSA, STS and ARSE.

Subunit:

Monomer, homodimer and heterodimer with SUMF2.

Subcellular Location:

Endoplasmic reticulum lumen.

Tissue Specificity:

Ubiquitous. Highly expressed in kidney, pancreas and liver. Detected at lower levels in leukocytes, lung, placenta, small intestine, skeletal muscle and heart.

Post-translational modifications:

N-glycosylated. Contains high-mannose-type oligosaccharides.

DISEASE:

Defects in SUMF1 are the cause of multiple sulfatase deficiency (MSD) [MIM:272200]. MSD is a clinically and biochemically heterogeneous disorder caused by the simultaneous impairment of all sulfatases, due to defective post-translational modification and activation. It combines features of individual sulfatase deficiencies such as metachromatic leukodystrophy, mucopolysaccharidosis, chondrodysplasia punctata, hydrocephalus, ichthyosis, neurologic deterioration and developmental delay. Inheritance is autosomal recessive.

Similarity:

Belongs to the sulfatase-modifying factor family.

SWISS:

Q8NBK3

Gene ID:

285362

Database links:

[Entrez Gene: 536435](#)Cow

[Entrez Gene: 484681](#)Dog

[Entrez Gene: 100734272](#)Guinea pig

[Entrez Gene: 100052857](#)Horse

[Entrez Gene: 285362](#)Human

[Entrez Gene: 58911](#)Mouse

[Entrez Gene: 100514188](#)Pig

[Entrez Gene: 362409](#)Rat

[Omim: 607939](#)Human

[SwissProt: Q0P5L5](#)Cow

[SwissProt: Q8NBK3](#)Human

[SwissProt: Q8R0F3](#)Mouse

Important Note:

This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.

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