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Rabbit Anti-FACE1/ZMPSTE24 antibody

SL4392R

Product Name:	FACE1/ZMPSTE24
Chinese Name:	锌金属蛋白酶STE24同源蛋白抗体
Alias:	CAAX prenyl protease 1 homolog; FACE-1; FACE1; FACE1_HUMAN; Farnesylated proteins converting enzyme 1; Farnesylated proteins-converting enzyme 1; Prenyl protein specific endoprotease 1; Prenyl protein-specific endoprotease 1; STE24; Zinc metalloproteinase Ste24 homolog; zmpste24.
Organism Species:	Rabbit
Clonality:	Polyclonal
React Species:	Human,Mouse,Rat,Chicken,Dog,Cow,Horse,Rabbit,Sheep,
Applications:	ELISA=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:	55kDa
Cellular localization:	The nucleuscytoplasmic
Form:	Lyophilized or Liquid
Concentration:	1mg/ml
immunogen:	KLH conjugated synthetic peptide derived from human FACE1/ZMPSTE24:381-475/475
Lsotype:	IgG
Purification:	affinity purified by Protein A
Storage Buffer:	Preservative: 15mM Sodium Azide, Constituents: 1% BSA, 0.01M PBS, pH 7.4
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:	This gene encodes a member of the peptidase M48A family. The encoded protein is a zinc metalloproteinase involved in the two step post-translational proteolytic cleavage of carboxy terminal residues of farnesylated prelamin A to form mature lamin A.

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Mutations in this gene have been associated with mandibuloacral dysplasia and restrictive dermopathy. [provided by RefSeq, Jul 2008]

Function:
Proteolytically removes the C-terminal three residues of farnesylated proteins. Acts on lamin A/C.

Subcellular Location:
Endoplasmic reticulum membrane. Golgi apparatus membrane.

Tissue Specificity:
Widely expressed. High levels in kidney, prostate, testis and ovary.

DISEASE:
Defects in ZMPSTE24 are the cause of mandibuloacral dysplasia with type B lipodystrophy (MADB) [MIM:608612]. Mandibuloacral dysplasia (MAD) is a rare autosomal recessive disorder characterized by mandibular and clavicular hypoplasia, acroosteolysis, delayed closure of the cranial suture, joint contractures, and types A or B patterns of lipodystrophy. Type B lipodystrophy observed in MADB, is characterized by generalized fat loss.
Defects in ZMPSTE24 are a cause of lethal tight skin contracture syndrome (LTSCS) [MIM:275210]; also called restrictive dermopathy (RD). Lethal tight skin contracture syndrome is a rare disorder mainly characterized by intrauterine growth retardation, tight and rigid skin with erosions, prominent superficial vasculature and epidermal hyperkeratosis, facial features (small mouth, small pinched nose and micrognathia), sparse/absent eyelashes and eyebrows, mineralization defects of the skull, thin dysplastic clavicles, pulmonary hypoplasia, multiple joint contractures and an early neonatal lethal course. Liveborn children usually die within the first week of life. The overall prevalence of consanguineous cases suggested an autosomal recessive inheritance.

Similarity:
Belongs to the peptidase M48A family.

SWISS:
O75844

Gene ID:
10269

Database links:

Entrez Gene: 10269 Human

Omim: 606480 Human

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	SwissProt: O75844 Human Unigene: 132642 Human Unigene: 721062 Human **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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