

Rabbit Anti-Nesprin 1/AF350 Conjugated antibody

SL19207R-AF350

Product Name	Anti-Nesprin 1/AF350
Chinese Name	AF350 标记的突触核膜蛋白 1 抗体
Alias	CPG2; Enaptin; Myne-1; MYNE1; Myocyte nuclear envelope protein 1; Nesprin-1; Nuclear envelope spectrin repeat protein 1; SCAR8; Synaptic nuclear envelope protein 1; Syne-1; SYNE1; SYNE1_HUMAN; SYNE1B.
Research Area	Cardiovascular Cell biology Neurobiology Cell differentiation
Immunogen Species	Rabbit
Clonality	Polyclonal
React Species	Rat(predicted:Human,Mouse,Dog,Cow,Horse,Sheep) IF=1:100-500
Applications	not yet tested in other applications. optimal dilutions/concentrations should be determined by the end user.
Molecular weight	1010kDa
Form	Lyophilized or Liquid
Concentration	1mg/ml
immunogen	KLH conjugated synthetic peptide derived from human Nesprin 1
Lsotype	IgG
Purification	affinity purified by Protein A
Storage Buffer	1M TBS(pH7.4) with 1% BSA, 3% Proclin300 and 50% Glycerol 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 1M PBS or diluent of antibody the antibody is stable for at least two weeks at 2-4 °C.
Storage	
Product Detail	background: This gene encodes a spectrin repeat containing protein expressed in skeletal and smooth muscle, and peripheral blood lymphocytes, that localizes to the nuclear membrane. Mutations in this gene have been associated with autosomal recessive spinocerebellar ataxia 8, also referred to as autosomal

recessive cerebellar ataxia type 1 or recessive ataxia of Beauce. Alternatively spliced transcript variants encoding different isoforms have been described. [provided by RefSeq, Jul 2008]

Function:

Multi-isomeric modular protein which forms a linking network between organelles and the actin cytoskeleton to maintain the subcellular spatial organization. Component of SUN-protein-containing multivariate complexes also called LINC complexes which link the nucleoskeleton and cytoskeleton by providing versatile outer nuclear membrane attachment sites for cytoskeletal filaments. Involved in the maintenance of nuclear organization and structural integrity. Connects nuclei to the cytoskeleton by interacting with the nuclear envelope and with F-actin in the cytoplasm. Required for centrosome migration to the apical cell surface during early ciliogenesis.

Subcellular Location:

Nucleus outer membrane. Cytoplasm, cytoskeleton. Cytoplasm, myofibril, sarcomere. The largest part of the protein is cytoplasmic, while its C-terminal part is associated with the nuclear envelope, most probably the outer nuclear membrane. In skeletal and smooth muscles, a significant amount is found in the sarcomeres.

Tissue Specificity:

Widely expressed. Highly expressed in skeletal and smooth muscles, heart, spleen, and peripheral blood leukocytes.

DISEASE:

Defects in SYNE1 are the cause of spinocerebellar ataxia autosomal recessive type 8 (SCAR8) [MIM:610743]; also known as autosomal recessive cerebellar ataxia type 1 (ARCA1) or recessive ataxia of Beauce. Spinocerebellar ataxia is a clinically and genetically heterogeneous group of cerebellar disorders. Patients show progressive incoordination of gait and often poor coordination of hands, speech and eye movements, due to degeneration of the cerebellum with variable involvement of the brainstem and spinal cord. SCAR8 is an autosomal recessive form.

Defects in SYNE1 are the cause of Emery-Dreifuss muscular dystrophy type 4 (EDMD4) [MIM:612998]. A degenerative myopathy characterized by weakness and atrophy of muscle without involvement of the nervous system, early contractures of the elbows, Achilles tendons and spine, and cardiomyopathy associated with cardiac conduction defects.

Similarity:

Belongs to the nesprin family.
Contains 1 actin-binding domain.

Contains 2 CH (calponin-homology) domains.

Contains 12 HAT repeats.

Contains 1 KASH domain.

Contains 31 spectrin repeats.

Database links:

[Entrez Gene: 23345](#) Human

[Entrez Gene: 64009](#) Mouse

[Entrez Gene: 499010](#) Rat

[Omim: 608441](#) Human

[SwissProt: Q8NF91](#) Human

[SwissProt: Q6ZWR6](#) Mouse

[Unigene: 12967](#) Human

[Unigene: 331626](#) Mouse

Important Note:

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