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Nesprin 1 Rabbit pAb

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Catalog # AP57437

Product Information

Application	IHC-P, IHC-F, IF
Primary Accession	Q8NF91
Reactivity	Rat
Predicted	Dog, Human, Mouse, Horse, Sheep
Host	Rabbit
Clonality	Polyclonal
Calculated MW	1011086
Physical State	Liquid
Immunogen	KLH conjugated synthetic peptide derived from human Nesprin 1
Epitope Specificity	51-150/8798
Isotype	IgG
Purity	affinity purified by Protein A
Buffer	0.01M TBS (pH7.4) with 1% BSA, 0.02% Proclin300 and 50% Glycerol.
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.
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.
Important Note	This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.
Background Descriptions	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]

Additional Information

Gene ID	23345
Other Names	Nesprin-1, Enaptin, KASH domain-containing protein 1, KASH1, Myocyte nuclear envelope protein 1, Myne-1, Nuclear envelope spectrin repeat protein 1, Synaptic nuclear envelope protein 1, Syne-1, SYNE1 (HGNC:17089)
Dilution	IHC-P=1:100-500,IHC-F=1:100-500,IF=1:100-500
Storage	Store at -20 °C for one year. Avoid repeated freeze/thaw cycles. 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.

Protein Information

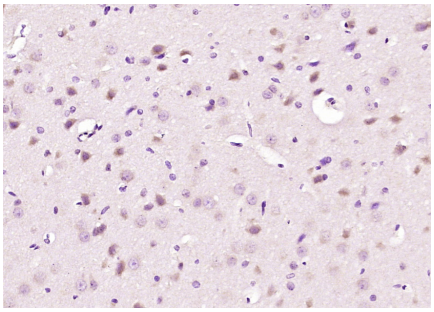
Name	SYNE1 (HGNC:17089)
Function	Multi-isomeric modular protein which forms a linking network between organelles and the actin cytoskeleton to maintain the subcellular spatial organization. As a component of the LINC (LInker of Nucleoskeleton and Cytoskeleton) complex involved in the connection between the nuclear lamina and the cytoskeleton. The nucleocytoplasmic interactions established by the LINC complex play an important role in the transmission of mechanical forces across the nuclear envelope and in nuclear movement and positioning. May be involved in nucleus- centrosome attachment and nuclear migration in neural progenitors implicating LINC complex association with SUN1/2 and probably association with cytoplasmic dynein-dynactin motor complexes; SYNE1 and SYNE2 may act redundantly. Required for centrosome migration to the apical cell surface during early ciliogenesis. May be involved in nuclear remodeling during sperm head formation in spermatogenesis; a probable SUN3:SYNE1/KASH1 LINC complex may tether spermatid nuclei to posterior cytoskeletal structures such as the manchette.
Cellular Location	Nucleus outer membrane; Single-pass type IV membrane protein; Cytoplasmic side Nucleus. Nucleus envelope. Cytoplasm, cytoskeleton. Cytoplasm, myofibril, sarcomere. Note=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. In myoblasts, relocalized from the nuclear envelope to the nucleus and cytoplasm during cell differentiation
Tissue Location	Expressed in HeLa, A431, A172 and HaCaT cells (at protein level). Widely expressed. Highly expressed in skeletal and smooth muscles, heart, spleen, peripheral blood leukocytes, pancreas, cerebellum, stomach, kidney and placenta. Isoform GSRP-56 is predominantly expressed in heart and skeletal muscle (at protein level).

Background

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Images

Paraformaldehyde-fixed, paraffin embedded (rat brain);



Antigen retrieval by boiling in sodium citrate buffer (pH6.0) for 15min; Block endogenous peroxidase by 3% hydrogen peroxide for 20 minutes; Blocking buffer (normal goat serum) at 37°C for 30min; Antibody incubation with (Nesprin 1) Polyclonal Antibody, Unconjugated (AP57437) at 1:200 overnight at 4°C, followed by operating according to SP Kit(Rabbit) (sp-0023) instructionsand DAB staining.

Please note: All products are 'FOR RESEARCH USE ONLY. NOT FOR USE IN DIAGNOSTIC OR THERAPEUTIC PROCEDURES'.