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# Product datasheet

# Recombinant Human Nesprin1/Syne-1 protein ab161609

# 1 Image

**Description** 

Product name Recombinant Human Nesprin1/Syne-1 protein

**Expression system** Wheat germ

Protein length Protein fragment

Animal free No

**Nature** Recombinant

**Species** Human

Sequence LRKIQQSVSEFEDKLAVPIKICSSATETYKVLQEHMDLCQA

LESLSSAIT

AFSASARKVVNRDSCVQEAAALQQQYEDILRRAKERQTA

LENLLAHWQRL EKELSSFLTW

Amino acids 1561 to 1670

Tags GST tag N-Terminus

#### **Specifications**

Our **Abpromise guarantee** covers the use of **ab161609** in the following tested applications.

The application notes include recommended starting dilutions; optimal dilutions/concentrations should be determined by the end user.

Applications ELISA

Western blot

Form Liquid

Additional notes This product was previously labelled as Nesprin 1.

# **Preparation and Storage**

Stability and Storage Shipped on dry ice. Upon delivery aliquot and store at -80°C. Avoid freeze / thaw cycles.

pH: 8.00

Constituents: 0.31% Glutathione, 0.79% Tris HCI

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#### **General Info**

#### **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.

#### **Tissue specificity**

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

#### Involvement in 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.

#### Sequence similarities

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.

#### **Domain**

The KASH domain, which contains a transmembrane domain, mediates the nuclear envelope targeting and is involved in the binding to SUN1 and SUN2 through recognition of their SUN domains.

### **Cellular localization**

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.

## **Images**

SDS-PAGE - Recombinant Human Nesprin1/Syne-1 protein (ab161609)

ab161609 on a 12.5% SDS-PAGE stained with Coomassie Blue.

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