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Product datasheet

Recombinant Human Progerin protein ab93918

1 Image

Description

Product name	Recombinant Human Progerin protein
Purity	> 90 % SDS-PAGE.
Expression system	Escherichia coli
Accession	P02545
Protein length	Full length protein
Animal free	No
Nature	Recombinant
Species	Human
Sequence	MAHHHHHHVGTGSNDDDDKSPDMETPSQRRATRSGAQ ASSTPLSPTRITR LQEKEDLQELNDRLAVYIDRVRSLETENAGLRLRITESEEV VSREVSGIK AAYEAELGDARKTLDSVAKERARLQLELSKVREEFKELK ARNTKKEGDLI AAQARLKDLEALLNSKEAALSTALSEKRTLEGELHDLRGQ VAKLEAALGE AKKQLQDEMLRRVDAENRLQTMKEELDFQKNIYSEELRE TKRRHETRLVE IDNGKQREFESRLADALQELRAQHEDQVEQYKKELEKTY SAKLDNARQSA ERNSNLVGAAHEELQQSRIRIDSLSAQLSQLQKQLAAKEA KLRDLEDSLA RERDTSRRLLAEKEREMAEMRARMQQQLDEYQELLDIKL ALDMEIHAYRK LLEGEEERLRLSPSPTSQRSRGRASSHSSQTQGGGSVTK KRKLESTESRS SFSQHARTSGRVAVEEVDEEGKFVRLRNKSNEDQSMGN WQIKRQNGDDPL LTYRFPPKFTLKAGQVVTIWAAGAGATHSPPTDLVWKAQ NTWGCGNSLRT ALINSTGEEVAMRKLVRSVTVVEDDEDEDGDDLLHHHHG SHCSSSGDPAE YNLRSRTVLCGTCGQPADKASASGSGAQSPQNCSIM

Amino acids	1 to 614
Tags	His tag N-Terminus , DDDDK tag N-Terminus

Specifications

Our **Abpromise guarantee** covers the use of **ab93918** in the following tested applications.

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

Applications	Western blot SDS-PAGE
Form	Liquid

Preparation and Storage

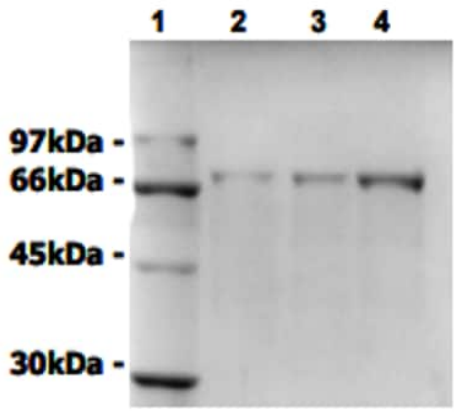
Stability and Storage	Shipped at 4°C. Upon delivery aliquot. Store at -80°C. Avoid freeze / thaw cycle. Constituents: 20% Glycerol, 0.315% Tris HCl, 2.922% Sodium chloride, 0.015% DTT, 0.044% EDTA

General Info

Function	Lamins are components of the nuclear lamina, a fibrous layer on the nucleoplasmic side of the inner nuclear membrane, which is thought to provide a framework for the nuclear envelope and may also interact with chromatin. Lamin A and C are present in equal amounts in the lamina of mammals. Plays an important role in nuclear assembly, chromatin organization, nuclear membrane and telomere dynamics. Required for normal development of peripheral nervous system and skeletal muscle and for muscle satellite cell proliferation. Required for osteoblastogenesis and bone formation. Also prevents fat infiltration of muscle and bone marrow, helping to maintain the volume and strength of skeletal muscle and bone. Prelamin-A/C can accelerate smooth muscle cell senescence. It acts to disrupt mitosis and induce DNA damage in vascular smooth muscle cells (VSMCs), leading to mitotic failure, genomic instability, and premature senescence.
Tissue specificity	In the arteries, prelamin-A/C accumulation is not observed in young healthy vessels but is prevalent in medial vascular smooth muscle cells (VSMCs) from aged individuals and in atherosclerotic lesions, where it often colocalizes with senescent and degenerate VSMCs. Prelamin-A/C expression increases with age and disease. In normal aging, the accumulation of prelamin-A/C is caused in part by the down-regulation of ZMPSTE24/FACE1 in response to oxidative stress.
Involvement in disease	Emery-Dreifuss muscular dystrophy 2, autosomal dominant Emery-Dreifuss muscular dystrophy 3, autosomal recessive Cardiomyopathy, dilated 1A Lipodystrophy, familial partial, 2 Limb-girdle muscular dystrophy 1B Charcot-Marie-Tooth disease 2B1 Hutchinson-Gilford progeria syndrome Cardiomyopathy, dilated, with hypergonadotropic hypogonadism Mandibuloacral dysplasia with type A lipodystrophy Lethal tight skin contracture syndrome Heart-hand syndrome Slovenian type

	Muscular dystrophy congenital LMNA-related Defects in LMNA may cause a late-onset cardiocutaneous progeria syndrome characterized by cutaneous manifestations of aging appearing in the third decade of life, cardiac valve calcification and dysfunction, prominent atherosclerosis, and cardiomyopathy, leading to death on average in the fourth decade.
Sequence similarities	Belongs to the intermediate filament family. Contains 1 LTD domain.
Post-translational modifications	Increased phosphorylation of the lamins occurs before envelope disintegration and probably plays a role in regulating lamin associations. Proteolytic cleavage of the C-terminal of 18 residues of prelamin-A/C results in the production of lamin-A/C. The prelamin-A/C maturation pathway includes farnesylation of CAAX motif, ZMPSTE24/FACE1 mediated cleavage of the last three amino acids, methylation of the C-terminal cysteine and endoproteolytic removal of the last 15 C-terminal amino acids. Proteolytic cleavage requires prior farnesylation and methylation, and absence of these blocks cleavage. Sumoylation is necessary for the localization to the nuclear envelope. Farnesylation of prelamin-A/C facilitates nuclear envelope targeting.
Cellular localization	Nucleus speckle and Nucleus. Nucleus envelope. Nucleus lamina. Nucleus, nucleoplasm. Farnesylation of prelamin-A/C facilitates nuclear envelope targeting and subsequent cleaveage by ZMPSTE24/FACE1 to remove the farnesyl group produces mature lamin-A/C, which can then be inserted into the nuclear lamina. EMD is required for proper localization of non-farnesylated prelamin-A/C.

Images



SDS-PAGE - Recombinant Human Progerin protein (ab93918)

Purified recombinant human progerin protein, ab93918 (lane 1, molecular weight standard; lane 2, 1µg; lane 3, 3µg; lane 4, 5µg) was separated by SDS-PAGE (10% polyacrylamide) and stained with Coomassie Blue.

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