

# Recombinant Rat Progerin protein ab93917

## 1 图像

### 描述

产品名称	重组大鼠Progerin蛋白
纯度	> 90 % SDS-PAGE.
表达系统	Escherichia coli
蛋白长度	Full length protein
无动物成分	No
性质	Recombinant
种属	Rat

### 技术指标

Our **Abpromise guarantee** covers the use of **ab93917** in the following tested applications.

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

应用	Western blot
	SDS-PAGE
形式	Liquid

### 制备和贮存

稳定性和存储	Shipped at 4°C. Upon delivery aliquot. Store at -80°C. Avoid freeze / thaw cycle.
	Constituent: 10% Glycerol

### 常规信息

功能	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,
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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.

#### 组织特异性

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.

#### 疾病相关

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.

#### 序列相似性

Belongs to the intermediate filament family.

Contains 1 LTD domain.

#### 翻译后修饰

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.

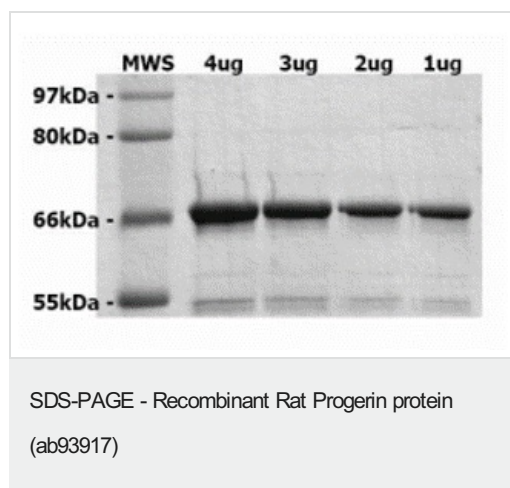
Farnesylation of prelamin-A/C facilitates nuclear envelope targeting.

#### 细胞定位

Nucleus speckle and Nucleus. Nucleus envelope. Nucleus lamina. Nucleus, nucleoplasm.

Farnesylation of prelamin-A/C facilitates nuclear envelope targeting and subsequent cleavage 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.

#### 图片



Purified recombinant full-length rat progerin protein, ab93917 was separated by SDS-PAGE (8% polyacrylamide) and stained with Coomassie Blue.

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