

Recombinant Human Lamin A protein ab83472

1 References 画像数 1

製品の詳細

製品名	Recombinant Human Lamin A protein
精製度	> 90 % SDS-PAGE.
発現系	Escherichia coli
アクセッション番号	<b>P02545-1</b>
タンパク質長	Full length protein
Animal free	No
由来	Recombinant
生物種	Human

配列	MAHHHHHHVGTGSNDDDDKSPDMETPSQRRATRSGAQASSTP LSPTRITR LQEKEDLQELNDR LAVYIDRVHSLETENAGLRLRITESEEVV SREVSGIK AAYEAE LGDARKTLD SVAKERARLQLELSKVREEFKELKARN TKKEGDLI AAQARLKDLEALLNSKEAALSTALSEKRTLEGELHDLRGQVA KLEAALGE AKKQLQDEMLRRVDAENRLQTMKEELDFQKNIYSEELRETKR RHETRLVE IDNGKQREFESRLADALQELRAQHEDQVEQYKKELEKTYSAK LDNARQSA ERNSNLVGAAHEELQQSRIRIDSLSAQLSQLQKQLAAKEAKL RDLED SLA RERDTSRLLAEKEREMAEMRARMQQQLDEYQELLDIKLALD MEIHAYRK LLEGEERLRLSPSPTSQRSRGRASSHSSQTQGGGSVTKKRK LESTESRS SFSQHARTSGRVAVEEVDEEGKFVRLRNKSNEDQSMGNWQIK RQNGDDPL LTYRFPPKFTLKAGQVVTIWAAGAGATHSPPTDLVWKAQNTW GCGNSLRT ALINSTGEEVAMRKLVR SVTVVEDDEDEDGDDLHHHHGSHC SSSGDPAE YNLRSRTVLCGTCGQPADKASASGSGAQVGGPISSGSSASSV
----	--

領域	1 to 645
タグ	His tag N-Terminus , DDDDK tag N-Terminus

## 特性

Our **Abpromise guarantee** covers the use of **ab83472** in the following tested applications.

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

アプリケーション	Western blot
	SDS-PAGE

製品の状態	Liquid
-------	--------

## 前処理および保存

保存方法および安定性	Shipped at 4°C. Upon delivery aliquot and store at -20°C. Avoid freeze / thaw cycles. pH: 7.00 Constituents: 20% Glycerol, 0.003% Sodium chloride, Phosphate Buffer, 0.015% DTT, 0.044% EDTA
------------	--

## 関連情報

機能	<p>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. Play an important role in nuclear assembly, chromatin organization, nuclear membrane and telomere dynamics.</p> <p>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.</p>
組織特異性	<p>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.</p>
関連疾患	<p>Defects in LMNA are the cause of Emery-Dreifuss muscular dystrophy type 2 (EDMD2) [MIM:181350]. 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.</p> <p>Defects in LMNA are the cause of cardiomyopathy dilated type 1A (CMD1A) [MIM:115200]. Dilated cardiomyopathy is a disorder characterized by ventricular dilation and impaired systolic function, resulting in congestive heart failure and arrhythmia. Patients are at risk of premature death.</p> <p>Defects in LMNA are the cause of familial partial lipodystrophy type 2 (FPLD2) [MIM:151660]; also known as familial partial lipodystrophy Dunnigan type. A disorder characterized by the loss of subcutaneous adipose tissue in the lower parts of the body (limbs, buttocks, trunk). It is accompanied by an accumulation of adipose tissue in the face and neck causing a double chin,</p>

fat neck, or cushingoid appearance. Adipose tissue may also accumulate in the axillae, back, labia majora, and intraabdominal region. Affected patients are insulin-resistant and may develop glucose intolerance and diabetes mellitus after age 20 years, hypertriglyceridemia, and low levels of high density lipoprotein cholesterol.

Defects in LMNA are the cause of limb-girdle muscular dystrophy type 1B (LGMD1B)

[MIM:159001]. LGMD1B is an autosomal dominant degenerative myopathy with age-related atrioventricular cardiac conduction disturbances, dilated cardiomyopathy, and the absence of early contractures. LGMD1B is characterized by slowly progressive skeletal muscle weakness of the hip and shoulder girdles. Muscle biopsy shows mild dystrophic changes.

Defects in LMNA are the cause of Charcot-Marie-Tooth disease type 2B1 (CMT2B1)

[MIM:605588]. CMT2B1 is a form of Charcot-Marie-Tooth disease, the most common inherited disorder of the peripheral nervous system. Charcot-Marie-Tooth disease is classified in two main groups on the basis of electrophysiologic properties and histopathology: primary peripheral demyelinating neuropathy or CMT1, and primary peripheral axonal neuropathy or CMT2.

Neuropathies of the CMT2 group are characterized by signs of axonal regeneration in the absence of obvious myelin alterations, normal or slightly reduced nerve conduction velocities, and progressive distal muscle weakness and atrophy. CMT2B1 inheritance is autosomal recessive.

Defects in LMNA are the cause of Hutchinson-Gilford progeria syndrome (HGPS) [MIM:176670].

HGPS is a rare genetic disorder characterized by features reminiscent of marked premature aging. Note=HGPS is caused by the toxic accumulation of a mutant form of lamin-A/C. This mutant protein, called progerin, acts to deregulate mitosis and DNA damage signaling, leading to premature cell death and senescence. Progerin lacks the conserved ZMPSTE24/FACE1 cleavage site and therefore remains permanently farnesylated. Thus, although it can enter the nucleus and associate with the nuclear envelope, it cannot incorporate normally into the nuclear lamina.

Defects in LMNA are the cause of cardiomyopathy dilated with hypergonadotropic hypogonadism (CMDHH) [MIM:212112]. A disorder characterized by the association of genital anomalies, hypergonadotropic hypogonadism and dilated cardiomyopathy. Patients can present other variable clinical manifestations including mental retardation, skeletal anomalies, scleroderma-like skin, graying and thinning of hair, osteoporosis. Dilated cardiomyopathy is characterized by ventricular dilation and impaired systolic function, resulting in congestive heart failure and arrhythmia.

Defects in LMNA are the cause of mandibuloacral dysplasia with type A lipodystrophy (MADA)

[MIM:248370]. A disorder characterized by mandibular and clavicular hypoplasia, acroosteolysis, delayed closure of the cranial suture, progeroid appearance, partial alopecia, soft tissue calcinosis, joint contractures, and partial lipodystrophy with loss of subcutaneous fat from the extremities. Adipose tissue in the face, neck and trunk is normal or increased.

Defects in LMNA are a cause of lethal tight skin contracture syndrome (LTSCS) [MIM:275210];

also known as restrictive dermopathy (RD). Lethal tight skin contracture syndrome is a rare disorder mainly characterized by intrauterine growth retardation, tight and rigid skin with erosions, prominent superficial vasculature and epidermal hyperkeratosis, facial features (small mouth, small pinched nose and micrognathia), sparse/absent eyelashes and eyebrows, mineralization defects of the skull, thin dysplastic clavicles, pulmonary hypoplasia, multiple joint contractures and an early neonatal lethal course. Liveborn children usually die within the first week of life. The overall prevalence of consanguineous cases suggested an autosomal recessive inheritance.

Defects in LMNA are the cause of heart-hand syndrome Slovenian type (HHS-Slovenian)

[MIM:610140]. Heart-hand syndrome (HHS) is a clinically and genetically heterogeneous disorder characterized by the co-occurrence of a congenital cardiac disease and limb malformations.

Defects in LMNA are the cause of muscular dystrophy congenital LMNA-related (CMD-LMNA)

[MIM:613205]. It is a form of congenital muscular dystrophy. Patients present at birth, or within the first few months of life, with hypotonia, muscle weakness and often with joint contractures.

## 配列類似性

Belongs to the intermediate filament family.

## 翻訳後修飾

Increased phosphorylation of the lamins occurs before envelope disintegration and probably plays a role in regulating lamin associations.

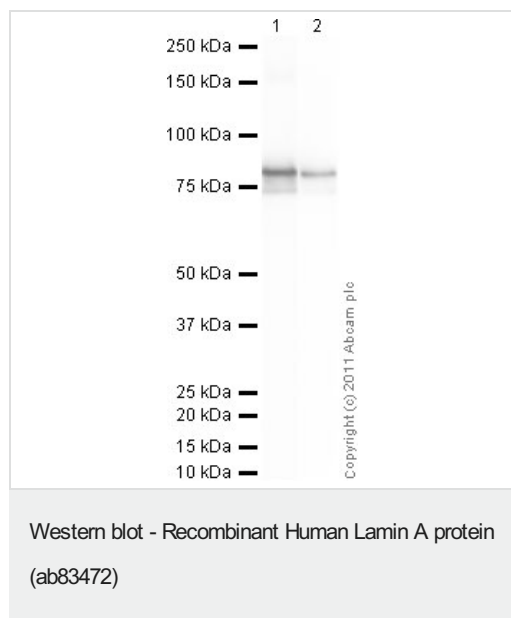
Proteolytic cleavage of the C-terminal of 18 residues of prelamin-A/C results in the production of lamin-A/C. The prelamin-A/C maturation pathway includes farnesylation of CAAX motif, ZMPSTE24/FACE1 mediated cleavage of the last three amino acids, methylation of the C-terminal cysteine and endoproteolytic 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.

## 細胞内局在

Nucleus. Nucleus envelope. 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.

## 画像



**All lanes :** Anti-Lamin A antibody ([ab26300](#)) at 1 µg/ml

**Lane 1 :** Recombinant Human Lamin A protein (ab83472) at 0.1 µg

**Lane 2 :** Recombinant Human Lamin A protein (ab83472) at 0.01 µg

### Secondary

**All lanes :** Goat Anti-Rabbit IgG H&L (HRP) preadsorbed ([ab97080](#)) at 1/5000 dilution

Developed using the ECL technique.

Performed under reducing conditions.

**Predicted band size:** 70 kDa

**Exposure time:** 10 seconds

**Please note:** All products are "FOR RESEARCH USE ONLY. NOT FOR USE IN DIAGNOSTIC PROCEDURES"

## Our Abpromise to you: Quality guaranteed and expert technical support

- Replacement or refund for products not performing as stated on the datasheet
- Valid for 12 months from date of delivery
- Response to your inquiry within 24 hours
- We provide support in Chinese, English, French, German, Japanese and Spanish

- Extensive multi-media technical resources to help you
- We investigate all quality concerns to ensure our products perform to the highest standards

If the product does not perform as described on this datasheet, we will offer a refund or replacement. For full details of the Abpromise, please visit <https://www.abcam.co.jp/abpromise> or contact our technical team.

#### **Terms and conditions**

---

- Guarantee only valid for products bought direct from Abcam or one of our authorized distributors