

Anti-Lamin A antibody [133A2] ab8980

★★★★★

17 Abreviews

87 References

8 图像

概述

产品名称	Anti-Lamin A抗体[133A2]
描述	小鼠单克隆抗体[133A2] to Lamin A
宿主	Mouse
特异性	Reacts against lamin A exclusively (the antibody was raised against the carboxy-terminus of 98 amino acids present in lamin A and absent from lamin C. As a result, this antibody recognizes lamin A but not lamin C; See Machiels et al 1997). Lamins do not appear to be universally distributed among different cell and tissue types. ab8980 has been tested in testis parenchyma and testicular germ cell tumours (See Machiels et al 1997). Other cell/tissue types have not been tested.
经测试应用	适用于: Flow Cyt, IHC-Fr, WB, IHC-P, ICC/IF
种属反应性	与反应: Mouse, Human
免疫原	Recombinant fragment corresponding to Human Lamin A (C terminal). Database link: P02545
表位	Recognizes an epitope located between residues 598-611 of lamin A.
阳性对照	Mouse heart normal tissue lysate - total protein (ab30291) can be used as a positive control in WB. This antibody gave a positive signal in the following cell types: HeLa WB: Wild-type HAP1 whole cell lysate; HeLa whole cell lysate
常规说明	<p>The Life Science industry has been in the grips of a reproducibility crisis for a number of years. Abcam is leading the way in addressing this with our range of recombinant monoclonal antibodies and knockout edited cell lines for gold-standard validation. Please check that this product meets your needs before purchasing.</p> <p>If you have any questions, special requirements or concerns, please send us an inquiry and/or contact our Support team ahead of purchase. Recommended alternatives for this product can be found below, along with publications, customer reviews and Q&As</p>

性能

形式	Liquid
存放说明	Shipped at 4°C. Store at +4°C short term (1-2 weeks). Upon delivery aliquot. Store at -20°C long term. Avoid freeze / thaw cycle.

存储溶液	Preservative: 0.09% Sodium azide Constituent: 99% PBS
纯度	This IgG3 antibody has to be purified from the culture medium by ion-exchange chromatography, therefore it will contain remnants of bovine serum albumin.
克隆	Ion Exchange Chromatography
克隆编号	单克隆
骨髓瘤	133A2
同种型	P3x63-Ag8.653
轻链类型	IgG3
	kappa

应用

The Abpromise guarantee **Abpromise™**承诺保证使用ab8980于以下的经测试应用

“应用说明”部分 下显示的仅为推荐的起始稀释度;实际最佳的稀释度/浓度应由使用者检定。

应用	Ab评论	说明
Flow Cyt		1/100 - 1/200. ab18392 - Mouse monoclonal IgG3, is suitable for use as an isotype control with this antibody.
IHC-Fr	★★★★★ (1)	1/100 - 1/200.
WB	★★★★★ (6)	1/100 - 1/1000. Predicted molecular weight: 74 kDa. PMID: 9274531
IHC-P	★★★★★ (4)	1/100 - 1/200.
ICC/IF	★★★★★ (6)	Use a concentration of 1 µg/ml.

靶标

功能	<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>
组织特异性	<p>In the arteries, prelamin-A/C accumulation is not observed in young healthy vessels but is prevalent in medial vascular smooth muscle celle (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</p>

oxidative stress.

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.

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.

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, 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.

序列相似性

Belongs to the intermediate filament family.

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.

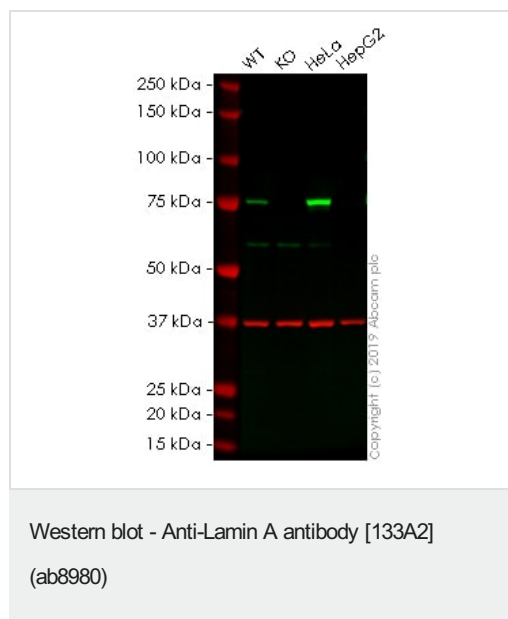
Sumoylation is necessary for the localization to the nuclear envelope.

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

细胞定位

Nucleus. 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.

图片



All lanes : Anti-Lamin A antibody [133A2] (ab8980) at 1/100 dilution

Lane 1 : Wild-type HAP1 whole cell lysate

Lane 2 : LMNA knockout HAP1 whole cell lysate

Lane 3 : HeLa whole cell lysate

Lane 4 : HepG2 whole cell lysate

Lysates/proteins at 20 µg per lane.

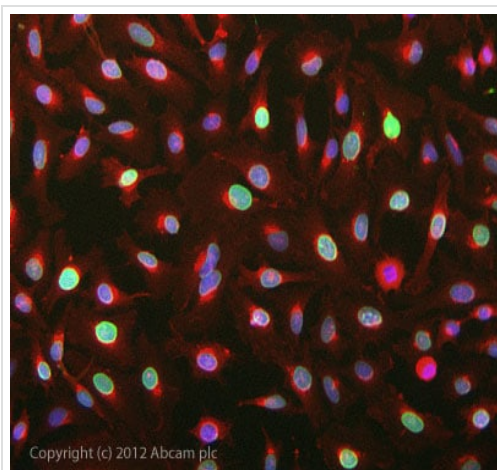
Predicted band size: 74 kDa

Observed band size: 74 kDa

Lanes 1 -4: Merged signal (red and green). Green - ab8980

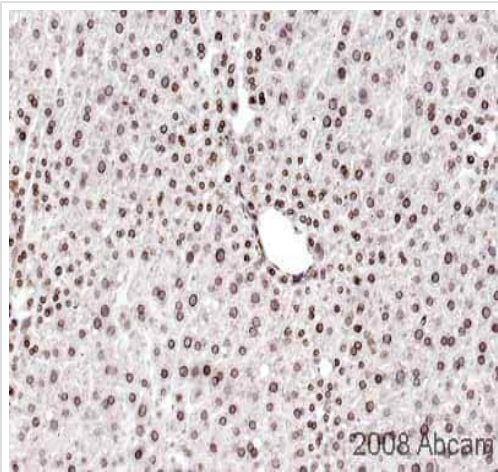
observed at 74 kDa. Red - loading control, **ab181602**, observed at 37 kDa.

ab8980 was shown to recognize in wild-type HAP1 cells as signal was lost at the expected MW in LMNA knockout cells. Additional cross-reactive bands were observed in the wild-type and knockout cells. Wild-type and LMNA knockout samples were subjected to SDS-PAGE. The membrane was blocked with 3pc Milk. Ab8980 and **ab181602** (Rabbit anti GAPDH loading control) were incubated overnight at 4°C at 1/100 dilution and 1/20000 dilution respectively. Blots were developed with Goat anti-Mouse IgG H&L (IRDye® 800CW) preabsorbed **ab216772** and Goat anti-Rabbit IgG H&L (IRDye® 680RD) preabsorbed **ab216777** secondary antibodies at 1/20000 dilution for 1 hour at room temperature before imaging.



ICC/IF image of ab8980 stained HeLa cells. The cells were 100% methanol fixed (5 min) and then incubated in 1%BSA / 10% normal goat serum (**ab7481**) / 0.3M glycine in 0.1% PBS-Tween for 1h to permeabilise the cells and block non-specific protein-protein interactions. The cells were then incubated with the antibody ab8980 at 1µg/ml overnight at +4°C. The secondary antibody (green) was DyLight® 488 goat anti- mouse (**ab96879**) IgG (H+L) used at a 1/1000 dilution for 1h. Alexa Fluor® 594 WGA was used to label plasma membranes (red) at a 1/200 dilution for 1h. DAPI was used to stain the cell nuclei (blue) at a concentration of 1.43µM.

Immunocytochemistry/ Immunofluorescence - Anti-Lamin A antibody [133A2] (ab8980)



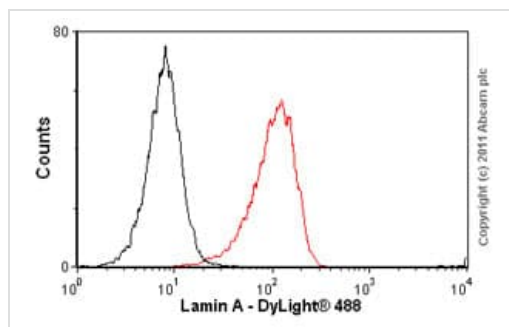
Immunohistochemistry (Formalin/PFA-fixed paraffin-embedded sections) - Anti-Lamin A antibody

[133A2] (ab8980)

This image is courtesy of an Abreview submitted by Mr Carl Hobbs

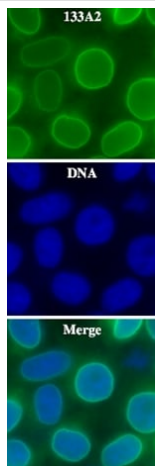
ab8980 staining of Lamin A in adult mouse liver sections.

Formaldehyde-fixed paraffin-embedded sections of mouse liver tissue were incubated with ab8980 (1/2500) for 2 hours. Antigen retrieval was performed by heat induction in citrate buffer pH 6.0 ([ab64214](#)). Please see accompanying abreview for additional information.



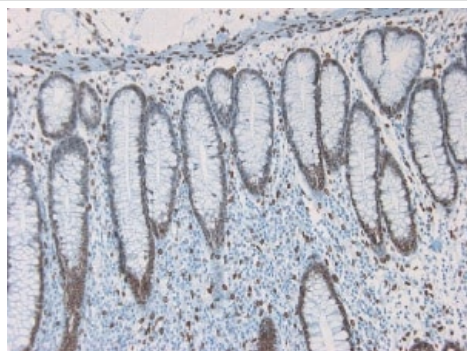
Flow Cytometry - Anti-Lamin A antibody [133A2] (ab8980)

Overlay histogram showing HeLa cells stained with ab8980 (red line). The cells were fixed with methanol (5 min) and then permeabilized with 0.1% PBS-Tween for 20 min. The cells were then incubated in 1x PBS / 10% normal goat serum / 0.3M glycine to block non-specific protein-protein interactions followed by the antibody (ab8980, 1/100 dilution) for 30 min at 22°C. The secondary antibody used was DyLight® 488 goat anti-mouse IgG (H+L) ([ab96879](#)) at 1/500 dilution for 30 min at 22°C. Isotype control antibody (black line) was mouse IgG3 [MG3-35] (2µg/1x10⁶ cells) used under the same conditions. Acquisition of >5,000 events was performed. This antibody gave a decreased signal in HeLa cells fixed with 4% paraformaldehyde (10 min)/permeabilized with 0.1% PBS-Tween used under the same conditions.



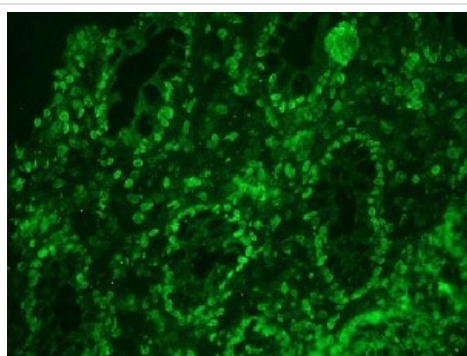
Immunocytochemistry/ Immunofluorescence - Anti-Lamin A antibody [133A2] (ab8980)

These images show HeLa cells stained with Laminin A antibody - ab8980 (green) and with DAPI (blue). The pictures were kindly supplied as part of the review submitted by Dr Josef Gotzmann at Medical University of Vienna, Austria.



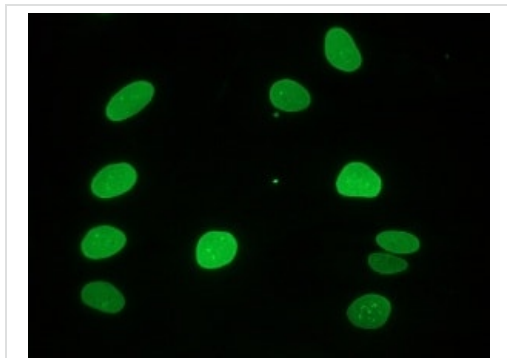
Immunohistochemistry (Formalin/PFA-fixed paraffin-embedded sections) - Anti-Lamin A antibody [133A2] (ab8980)

Immunohistochemistry on paraffin section of human colon.



Immunohistochemistry (Frozen sections) - Anti-Lamin A antibody [133A2] (ab8980)

Immunohistochemistry on frozen sections of human colon showing nuclear lamina staining in epithelial and connective tissue cells.



Immunocytochemical staining of fibroblasts showing nuclear lamina

Immunocytochemistry/ Immunofluorescence - Anti-Lamin A antibody [133A2] (ab8980)

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