abcam

Product datasheet

Anti-Lamin A + Lamin C antibody [JOL2] ab40567

KO VALIDATED

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Overview

Product name	Anti-Lamin A + Lamin C antibody [JOL2]
Description	Mouse monoclonal [JOL2] to Lamin A + Lamin C
Host species	Mouse
Specificity	This antibody reacts with both recombinant and native Lamin A and C in humans.
Tested applications	**Suitable for:** IHC-P, WB, Flow Cyt
Species reactivity	**Reacts with:** Human, African green monkey
Immunogen	Recombinant fragment corresponding to Human Lamin A + Lamin C.
Epitope	This product has been shown to bind to an epitope between amino acids 464-572.
Positive control	WB: HAP1, HeLa and HepG2 cell lysate IHC: Human skin and colon Tissue Flow Cyt: HeLa cells ICC/IF: HeLa and African Green Monkey CV1 cells.
General notes	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. 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

Properties

Form	Liquid
Storage instructions	Shipped at 4°C. Store at +4°C short term (1-2 weeks). Upon delivery aliquot. Store at -20°C long term.
Storage buffer	pH: 7.40 Preservative: 0.09% Sodium azide Constituents: 3.16% Tris HCl, Fetal calf serum
Purity	Tissue culture supernatant
Purification notes	This product is unpurified.
Clonality	Monoclonal

Clone number	JOL2
Myeloma	Sp2/0-Ag14
Isotype	IgG1

Applications

The Abpromise guarantee Our **Abpromise guarantee** covers the use of ab40567 in the following tested applications.

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

Application	Abreviews	Notes
IHC-P		Use at an assay dependent concentration. Perform antigen retrieval with 0.01 M sodium citrate buffer, pH 6.0 at 99-100°C for 20 minutes.
WB	★★★★☆ **(4)**	1/200. Predicted molecular weight: 74 kDa.
Flow Cyt		1/10. **ab170190** - Mouse monoclonal IgG1, is suitable for use as an isotype control with this antibody.

Target

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. Play an important role in nuclear assembly, chromatin organization, nuclear membrane and telomere dynamics.
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.

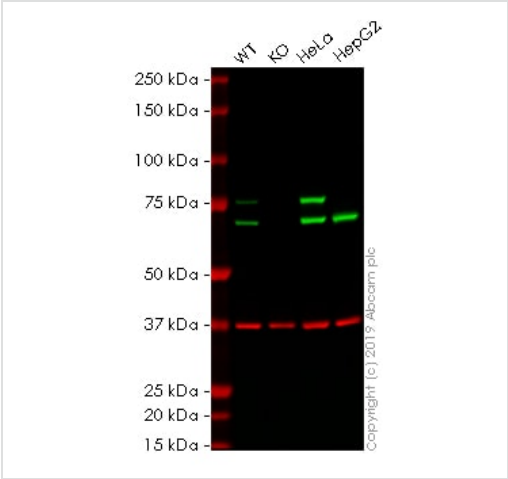
Tissue specificity In the arteries, prelamin-A/C accumulation is not observed in young healthy vessels but is prevalent in medial vascular smooth muscle celle (VSMCs) from aged individuals and in atherosclerotic lesions, where it often colocalizes with senescent and degenerate VSMCs. Prelamin-A/C expression 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 oxidative stress.

Involvement in disease 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, progeroide 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.

Sequence similarities	Belongs to the intermediate filament family.
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. Nucleus envelope. 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



Western blot - Anti-Lamin A + Lamin C antibody [JOL2] (ab40567)

All lanes : Anti-Lamin A + Lamin C antibody [JOL2] (ab40567) at 1/200 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

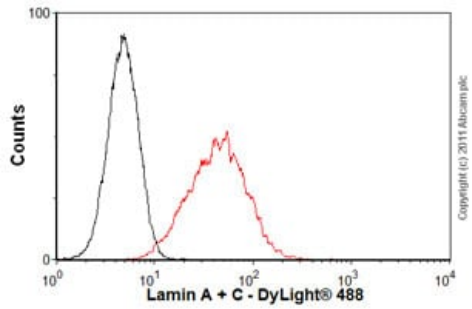
Lysates/proteins at 20 µg per lane.

Predicted band size: 74 kDa
Observed band size: 74 kDa

Lanes 1 - 4: Merged signal (red and green). Green - ab40567 observed at 74 kDa. Red - loading control, **ab181602**, observed at 37 kDa.

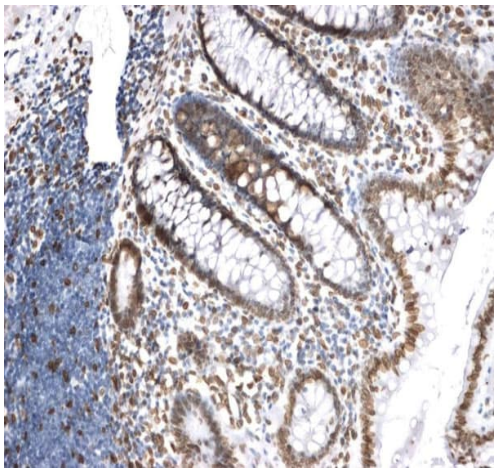
ab40567 was shown to recognize in wild-type HAP1 cells as signal was lost at the expected MW in LMNA knockout cells. Additional cross-reactive bands were observed in the wild-type and knockout cells. Wild-type and LMNA knockout samples were subjected to SDS-PAGE. The membrane was blocked with 3pc Milk. Ab40567 and **ab181602** (Rabbit anti GAPDH loading control) were incubated overnight at 4°C at 1/200 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.



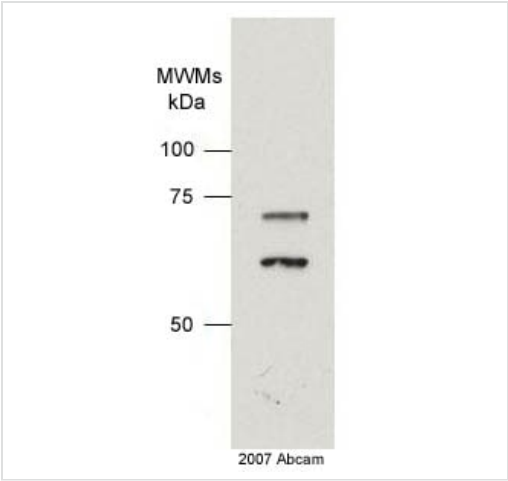
Flow Cytometry - Anti-Lamin A + Lamin C antibody [JOL2] (ab40567)

Overlay histogram showing HeLa cells stained with ab40567 (red line). The cells were fixed with 80% 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 (ab40567, 1/10 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 IgG1 [ICIGG1](**ab91353**, 2µg/1x10^6 cells) used under the same conditions. Acquisition of >5,000 events was performed.



Immunohistochemistry (Formalin/PFA-fixed paraffin-embedded sections) - Anti-Lamin A + Lamin C antibody [JOL2] (ab40567)

Immunohistochemistry (Formalin/PFA-fixed paraffin-embedded sections) analysis of human colon tissue sections labelling Lamin A + C with ab40567.



Western blot - Anti-Lamin A + Lamin C antibody [JOL2] (ab40567)

This image is courtesy of an anonymous Abreview

Anti-Lamin A + Lamin C antibody [JOL2] (ab40567) at 1/200 dilution + HeLa whole cell lysate (10ug)

Secondary

HRP conjugated goat anti-mouse antibody

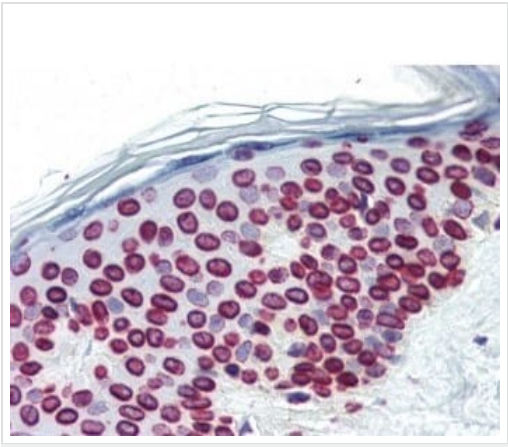
Developed using the ECL technique.

Performed under non-reducing conditions.

Predicted band size: 74 kDa

Observed band size: 64,73 kDa

Exposure time: 10 seconds



Immunohistochemistry (Formalin/PFA-fixed paraffin-embedded sections) - Anti-Lamin A + Lamin C antibody [JOL2] (ab40567)

Formalin-fixed, paraffin-embedded human skin tissue stained for Lamin A + C using ab40567 in immunohistochemical analysis. Slides were steamed in 0.01 M sodium citrate buffer, pH 6.0 at 99-100°C for 20 minutes and then let to stand at room temperature in a buffer for a further 20 minutes. They were then rinsed in 1X TBS with Tween (TBST) for 1 minute at room temperature.

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