

## Product datasheet

# Anti-Lamin A + Lamin C antibody [EPR4068] (Alexa Fluor® 647) ab223943

Recombinant RabMAb

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### Overview

<b>Product name</b>	Anti-Lamin A + Lamin C antibody [EPR4068] (Alexa Fluor® 647)
<b>Description</b>	Rabbit monoclonal [EPR4068] to Lamin A + Lamin C (Alexa Fluor® 647)
<b>Host species</b>	Rabbit
<b>Conjugation</b>	Alexa Fluor® 647. Ex: 652nm, Em: 668nm
<b>Specificity</b>	The antibody recognizes full length Lamin A/C and the cleaved small unit.
<b>Tested applications</b>	<b>Suitable for:</b> ICC/IF, Flow Cyt
<b>Species reactivity</b>	<b>Reacts with:</b> Human <b>Predicted to work with:</b> Mouse, Rat 
<b>Immunogen</b>	Synthetic peptide within Human Lamin A + Lamin C aa 1-100. The exact sequence is proprietary. Database link: <a href="#">P02545</a>
<b>Positive control</b>	ICC/IF: HeLa cells. Flow Cyt: HeLa cells
<b>General notes</b>	<p>Our RabMAb® technology is a patented hybridoma-based technology for making rabbit monoclonal antibodies. For details on our patents, please refer to <a href="#">RabMab® patents</a>.</p> <p>Alexa Fluor® is a registered trademark of Molecular Probes, Inc, a Thermo Fisher Scientific Company. The Alexa Fluor® dye included in this product is provided under an intellectual property license from Life Technologies Corporation. As this product contains the Alexa Fluor® dye, the purchase of this product conveys to the buyer the non-transferable right to use the purchased product and components of the product only in research conducted by the buyer (whether the buyer is an academic or for-profit entity). As this product contains the Alexa Fluor® dye the sale of this product is expressly conditioned on the buyer not using the product or its components, or any materials made using the product or its components, in any activity to generate revenue, which may include, but is not limited to use of the product or its components: in manufacturing; (ii) to provide a service, information, or data in return for payment (iii) for therapeutic, diagnostic or prophylactic purposes; or (iv) for resale, regardless of whether they are sold for use in research. For information on purchasing a license to this product for purposes other than research, contact Life Technologies Corporation, 5781 Van Allen Way, Carlsbad, CA 92008 USA or <a href="mailto:outlicensing@thermofisher.com">outlicensing@thermofisher.com</a>.</p> <p>This product is a <a href="#">recombinant rabbit monoclonal antibody</a>.</p>

## Properties

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<b>Form</b>	Liquid
<b>Storage instructions</b>	Shipped at 4°C. Store at +4°C short term (1-2 weeks). Upon delivery aliquot. Store at -20°C. Stable for 12 months at -20°C. Store In the Dark.
<b>Storage buffer</b>	pH: 7.40 Preservative: 0.02% Sodium azide Constituents: 30% Glycerol, 1% BSA, PBS
<b>Purity</b>	Protein A purified
<b>Clonality</b>	Monoclonal
<b>Clone number</b>	EPR4068
<b>Isotype</b>	IgG

## Applications

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Our [Abpromise guarantee](#) covers the use of **ab223943** 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
ICC/IF		1/1000. This product gave a positive signal in HeLa cells fixed with 100% methanol (5 min)
Flow Cyt		1/5000.

## Target

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<b>Function</b>	<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>
<b>Tissue specificity</b>	<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>
<b>Involvement in disease</b>	<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</p>

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.

### 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.

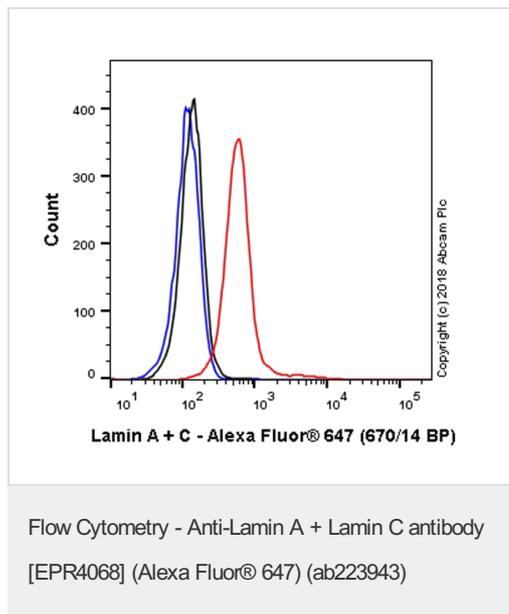
Proteolytic cleavage of the C-terminal of 18 residues of prelamin-A/C results in the production of lamin-A/C. The prelamin-A/C maturation pathway includes farnesylation of CAAX motif, ZMPSTE24/FACE1 mediated cleavage of the last three amino acids, methylation of the C-terminal cysteine and endoproteolytic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

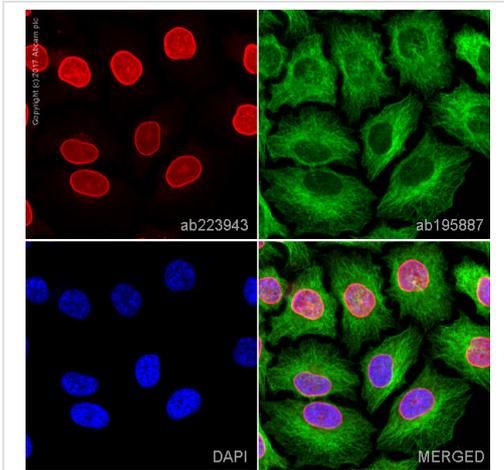
## Images



Overlay histogram showing HeLa cells stained with ab223943 (red line). The cells were fixed with 4% formaldehyde (10 min) and then permeabilized with 0.1% PBS-Triton X-100 for 15 min. The cells were then incubated in 1x PBS / 10% normal goat serum to block non-specific protein-protein interactions followed by the antibody (ab223943, 1/5000 dilution) for 30 min at 22°C.

Isotype control antibody (black line) was Rabbit IgG (monoclonal) Alexa Fluor® 647 (ab199093) used at the same concentration and conditions as the primary antibody. Unlabelled sample (blue line) was also used as a control.

Acquisition of >5,000 events were collected using a 40 mW Red laser (640nm) and 670/14 bandpass filter.



Immunocytochemistry/ Immunofluorescence - Anti-Lamin A + Lamin C antibody [EPR4068] (Alexa Fluor® 647) (ab223943)

ab223943 staining Lamin A + C in HeLa cells. The cells were fixed with 100% methanol (5 min), permeabilized with 0.1% Triton X-100 for 5 minutes and then blocked with 1% BSA/10% normal goat serum/0.3M glycine in 0.1% PBS-Tween for 1h. The cells were then incubated overnight at +4°C with ab223943 at 1/1000 dilution (shown in red) and ab195887, Mouse monoclonal to alpha Tubulin (Alexa Fluor® 488), at 1/250 dilution (shown in green). Nuclear DNA was labelled with DAPI (shown in blue). Image was taken with a confocal microscope (Leica-Microsystems, TCS SP8).

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