

PE Anti-Lamin A + Lamin C antibody [EPR4100] - Nuclear Envelope Marker ab210433

重组 RabMAB

3 图像

概述

产品名称	PE Anti-Lamin A + Lamin C抗体[EPR4100] -核Envelope Marker
描述	PE兔单克隆抗体[EPR4100] to Lamin A + Lamin C -核Envelope Marker
宿主	Rabbit
偶联物	PE. Ex: 488nm, Em: 575nm
特异性	The antibody recognizes full length Lamin A/C and the cleaved large unit.
经测试应用	适用于: ICC/IF, Flow Cyt (Intra)
种属反应性	与反应: Human
免疫原	Synthetic peptide. This information is proprietary to Abcam and/or its suppliers.
阳性对照	Flow Cyt (intra): HeLa cells ICC/IF: HeLa cells
常规说明	<p>This product is a recombinant monoclonal antibody, which offers several advantages including:</p> <ul style="list-style-type: none">- High batch-to-batch consistency and reproducibility- Improved sensitivity and specificity- Long-term security of supply- Animal-free production <p>For more information see here.</p> <p>Our RabMAB[®] technology is a patented hybridoma-based technology for making rabbit monoclonal antibodies. For details on our patents, please refer to RabMAB[®] patents.</p>

性能

形式	Liquid
存放说明	Shipped at 4°C. Upon delivery aliquot. Store at +4°C. Do Not Freeze. Store In the Dark.
存储溶液	pH: 7.4 Preservative: 0.02% Sodium azide Constituents: PBS, 1% BSA
纯度	Protein A purified
克隆	单克隆

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.

Belongs to the intermediate filament family.

Increased phosphorylation of the lamins occurs before envelope disintegration and probably plays

序列相似性

翻译后修饰

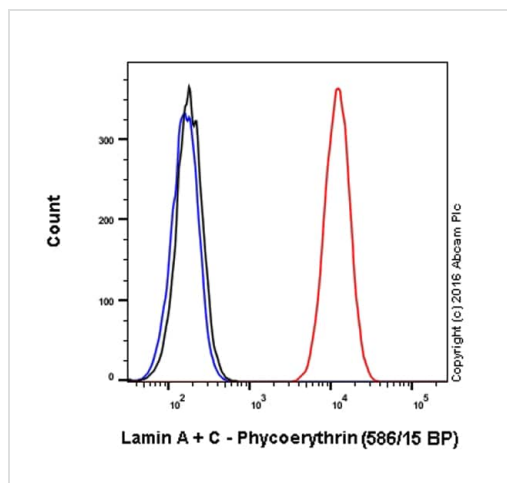
a role in regulating lamin associations.

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

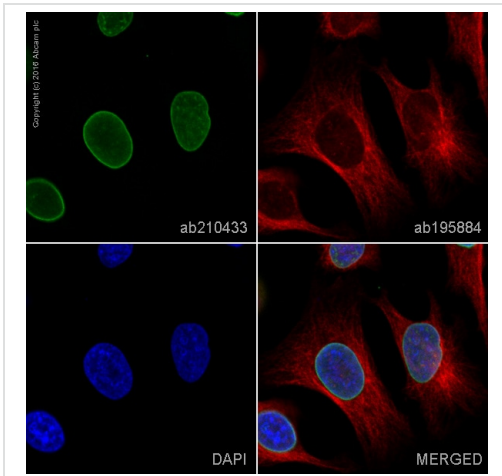


Flow Cytometry (Intracellular) - PE Anti-Lamin A + Lamin C antibody [EPR4100] - Nuclear Envelope Marker (ab210433)

Overlay histogram showing HeLa cells stained with ab210433 (red line). The cells were fixed with 4% formaldehyde (10 min) and then permeabilized with 90% methanol for 30 min at -20°C. The cells were then incubated in 1x PBS / 10% normal goat serum to block non-specific protein-protein interactions followed by the antibody (ab210433, 1/5000 dilution) for 30 min at 22°C.

Isotype control antibody (black line) was rabbit IgG (monoclonal) Phycoerythrin ([ab209478](#)) 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 50mW Yellow-Green laser (561nm) and 586/15 bandpass filter.







ab210433 staining Lamin A + C in HeLa cells. The cells were fixed with 100% methanol (5min), 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 ab210433 at 1/1000 dilution (**pseudocolored in green**) and **ab195884**, Rat monoclonal to Tubulin (Alexa Fluor® 647), at 1/250 dilution (shown in red). Nuclear DNA was labelled with DAPI (shown in blue).

Image was taken with a confocal microscope (Leica-Microsystems, TCS SP8).

This product also gave a positive signal under the same testing conditions in HeLa cells fixed with 4% formaldehyde (10 min).

Immunocytochemistry/ Immunofluorescence - PE
Anti-Lamin A + Lamin C antibody [EPR4100] -
Nuclear Envelope Marker (ab210433)

Why choose a recombinant antibody?

 Research with confidence Consistent and reproducible results	 Long-term and scalable supply Recombinant technology
 Success from the first experiment Confirmed specificity	 Ethical standards compliant Animal-free production

PE Anti-Lamin A + Lamin C antibody [EPR4100] -
Nuclear Envelope Marker (ab210433)

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

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