

Anti-Lamin A + Lamin C antibody [EPR4519(2)] ab169532

KO 評価済 リコンビナント RabMAb

★★★★★ [1 Abreviews](#) [9 References](#) [画像数 10](#)

製品の概要

製品名	Anti-Lamin A + Lamin C antibody [EPR4519(2)]
製品の詳細	Rabbit monoclonal [EPR4519(2)] to Lamin A + Lamin C
由来種	Rabbit
特異性	ab169532 recognizes full length Lamin A + C and cleaved large unit.
アプリケーション	適用あり: WB, IHC-P 適用なし: Flow Cyt, ICC/IF or IP
種交差性	交差種: Mouse, Rat, Human
免疫原	Synthetic peptide corresponding to Human Lamin A + Lamin C.
ポジティブ・コントロール	WB: HAP1, HepG2, PC-12, THP-1, HeLa + Staurosporine and HeLa cell lysates. IHC-P: Human skin and Human uterus tissues.
特記事項	This product is a recombinant monoclonal antibody, which offers several advantages including: <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 For more information see here . Our RabMAb [®] technology is a patented hybridoma-based technology for making rabbit monoclonal antibodies. For details on our patents, please refer to RabMAb[®] patents .

製品の特性

製品の状態	Liquid
保存方法	Shipped at 4°C. Store at +4°C short term (1-2 weeks). Upon delivery aliquot. Store at -20°C long term.
バッファー	pH: 7.2 Preservative: 0.01% Sodium azide Constituents: 9% PBS, 40% Glycerol (glycerin, glycerine), 0.05% BSA, 50% Tissue culture supernatant
精製度	Tissue culture supernatant
ポリ/モノ	モノクローナル

クローン名	EPR4519(2)
アイソタイプ	IgG

アプリケーション

The Abpromise guarantee **Abpromise保証は、** 次のテスト済みアプリケーションにおけるab169532の使用に適用されます
アプリケーションノートには、推奨の開始希釈率がありますが、適切な希釈率につきましてはご確認ください。

アプリケーション	Abreviews	特記事項
WB	★★★★★ (1)	1/10000 - 1/50000. Predicted molecular weight: 74 kDa.
IHC-P		1/250 - 1/500. Perform heat mediated antigen retrieval with citrate buffer pH 6 before commencing with IHC staining protocol.

追加情報 Is unsuitable for Flow Cyt, ICC/IF or IP.

ターゲット情報

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, 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</p>

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 a role in regulating lamin associations.

配列類似性

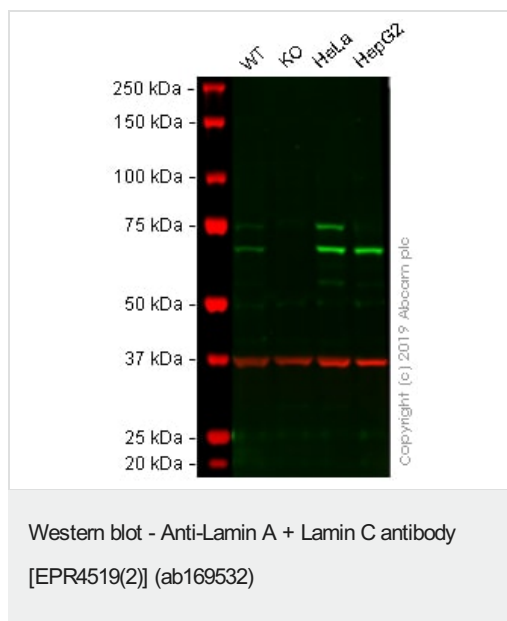
翻訳後修飾

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 + Lamin C antibody [EPR4519(2)] (ab169532) at 1/5000 dilution

Lane 1 : Wild-type HAP1 cell lysate

Lane 2 : LMNA knockout HAP1 cell lysate

Lane 3 : HeLa cell lysate

Lane 4 : HepG2 cell lysate

Lysates/proteins at 20 µg per lane.

Performed under reducing conditions.

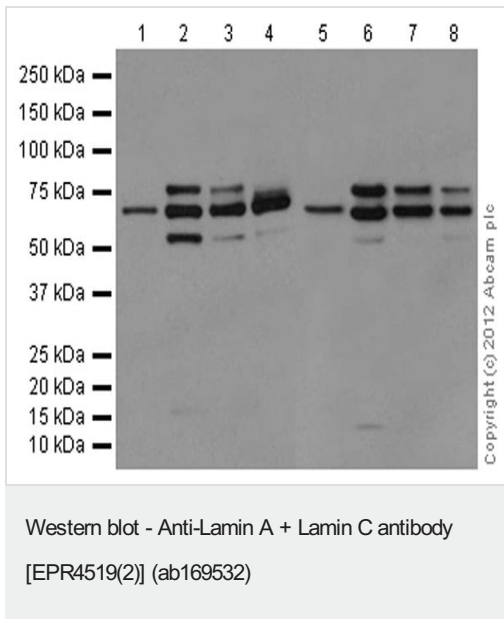
Predicted band size: 74 kDa

Observed band size: 70-75 kDa

Lanes 1 - 4: Merged signal (red and green). Green - ab169532 observed at 70-75 kDa. Red - loading control, [ab8245](#) (Mouse anti-GAPDH antibody [6C5]) observed at 37kDa.

ab169532 was shown to react with LMNA in wild-type HAP1 cells in Western blot. Loss of signal was observed when LMNA knockout sample was used. HAP1 wild-type and LMNA knockout cell lysates were subjected to SDS-PAGE. Membranes were blocked in 3% Milk in TBS-T (0.1% Tween®) before incubation with ab169532 and [ab8245](#) (Mouse anti-GAPDH antibody [6C5]) overnight at 4°C at a 1 in 5000 dilution and a 1 in 20000 dilution respectively. Blots were developed with Goat anti-Rabbit IgG H&L (IRDye® 800CW) preabsorbed ([ab216773](#)) and Goat anti-Mouse IgG H&L (IRDye® 680RD) preabsorbed ([ab216776](#)) secondary antibodies at 1 in

20000 dilution for 1 hour at room temperature before imaging.



All lanes : Anti-Lamin A + Lamin C antibody [EPR4519(2)]
(ab169532) at 1/5000 dilution

Lane 1 : Mouse brain tissue lysate

Lane 2 : Mouse heart tissue lysate

Lane 3 : Mouse kidney tissue lysate

Lane 4 : Mouse spleen tissue lysate

Lane 5 : Rat brain tissue lysate

Lane 6 : Rat heart tissue lysate

Lane 7 : Rat kidney tissue lysate

Lane 8 : Rat spleen tissue lysate

Lysates/proteins at 10 µg per lane.

Secondary

All lanes : Goat Anti-Rabbit IgG, (H+L), Peroxidase conjugated at
1/2000 dilution

Predicted band size: 74 kDa

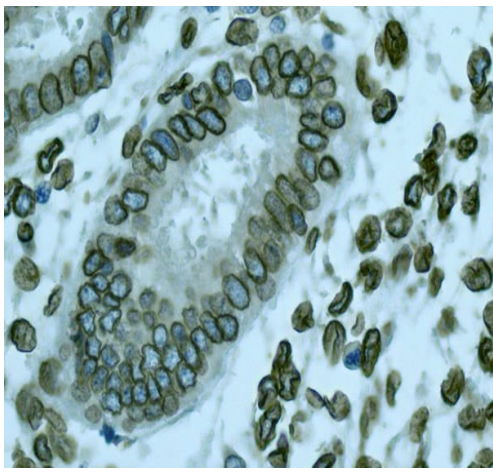
Observed band size: 74 kDa

Exposure time: 30 seconds

Blocking and Diluting buffer 5% NFDM /TBST

65-74 kDa: full length;

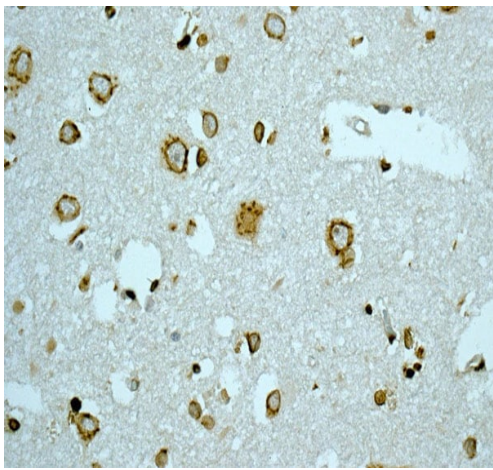
45-50 kDa: cleavage forms



Immunohistochemistry (Formalin/PFA-fixed paraffin-embedded sections) - Anti-Lamin A + Lamin C antibody [EPR4519(2)] (ab169532)

ab169532 showing +ve staining in Human normal colon tissue.

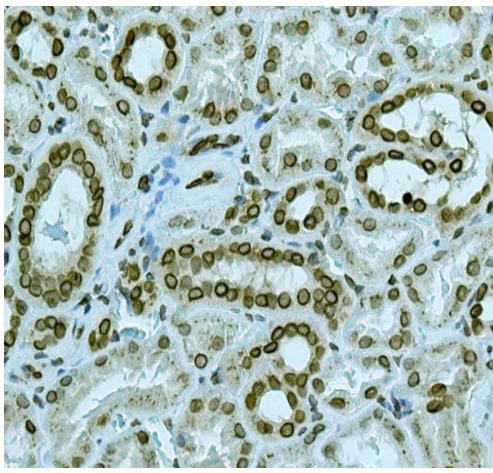
Perform heat mediated antigen retrieval with citrate buffer pH 6 before commencing with IHC staining protocol.



Immunohistochemistry (Formalin/PFA-fixed paraffin-embedded sections) - Anti-Lamin A + Lamin C antibody [EPR4519(2)] (ab169532)

ab169532 showing +ve staining in Human normal brain tissue.

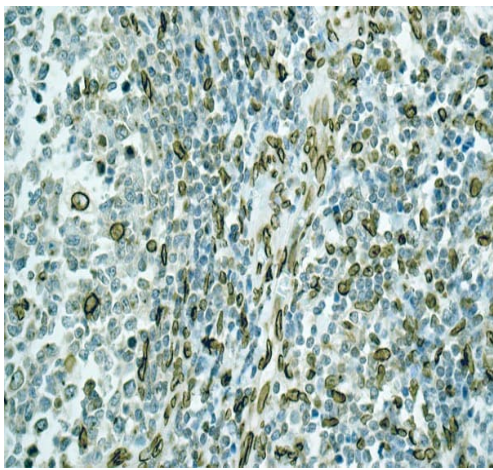
Perform heat mediated antigen retrieval with citrate buffer pH 6 before commencing with IHC staining protocol.



Immunohistochemistry (Formalin/PFA-fixed paraffin-embedded sections) - Anti-Lamin A + Lamin C antibody [EPR4519(2)] (ab169532)

ab169532 showing +ve staining in Human normal kidney tissue.

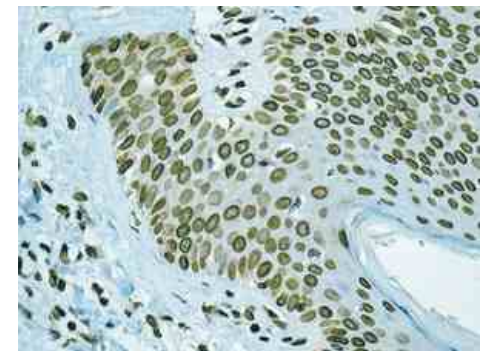
Perform heat mediated antigen retrieval with citrate buffer pH 6 before commencing with IHC staining protocol.



Immunohistochemistry (Formalin/PFA-fixed paraffin-embedded sections) - Anti-Lamin A + Lamin C antibody [EPR4519(2)] (ab169532)

ab169532 showing +ve staining in Human normal tonsil tissue.

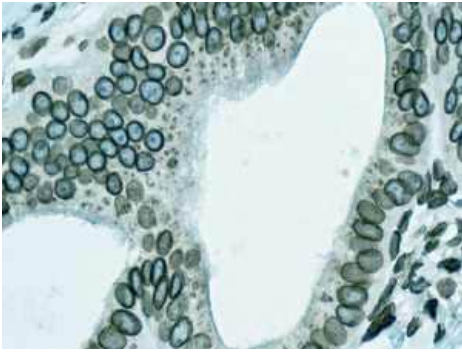
Perform heat mediated antigen retrieval with citrate buffer pH 6 before commencing with IHC staining protocol.



Immunohistochemistry (Formalin/PFA-fixed paraffin-embedded sections) - Anti-Lamin A + Lamin C antibody [EPR4519(2)] (ab169532)

Immunohistochemical analysis of paraffin-embedded Human skin tissue labeling Lamin A + C with ab169532 at 1/250 dilution.

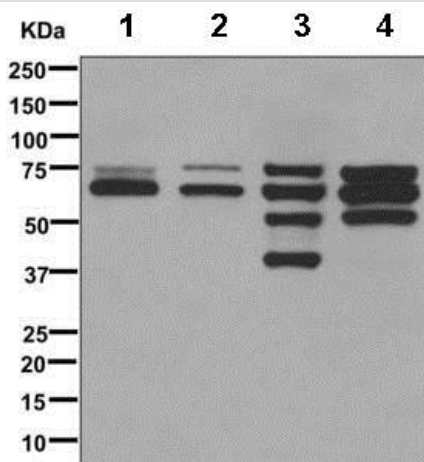
Perform heat mediated antigen retrieval with citrate buffer pH 6 before commencing with IHC staining protocol.



Immunohistochemistry (Formalin/PFA-fixed paraffin-embedded sections) - Anti-Lamin A + Lamin C antibody [EPR4519(2)] (ab169532)

Immunohistochemical analysis of paraffin-embedded Human uterus tissue labeling Lamin A + C with ab169532 at 1/250 dilution.

Perform heat mediated antigen retrieval with citrate buffer pH 6 before commencing with IHC staining protocol.



Western blot - Anti-Lamin A + Lamin C antibody [EPR4519(2)] (ab169532)

All lanes : Anti-Lamin A + Lamin C antibody [EPR4519(2)] (ab169532) at 1/10000 dilution

Lane 1 : PC-12 cell lysate

Lane 2 : THP-1 cell lysate

Lane 3 : HeLa + Staurosporine cell lysate

Lane 4 : HeLa cell lysate

Lysates/proteins at 10 µg per lane.

Secondary

All lanes : HRP labeled goat anti-rabbit at 1/2000 dilution

Predicted band size: 74 kDa

Why choose a recombinant antibody?



Anti-Lamin A + Lamin C antibody [EPR4519(2)] (ab169532)

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