# abcam

## Product datasheet

# Alexa Fluor® 647 Anti-Lamin B Receptor/LBR antibody [E398L] ab201349

יובעדער RabMAb

画像数 2

#### 製品の概要

製品名 Alexa Fluor® 647 Anti-Lamin B Receptor/LBR antibody [E398L]

製品の詳細 Alexa Fluor® 647 Rabbit monoclonal [E398L] to Lamin B Receptor/LBR

由来種 Rabbit

標識 Alexa Fluor® 647. Ex: 652nm, Em: 668nm

アプリケーション 適用あり: ICC/IF 種交差性 交差種: Human

免疫原 Synthetic peptide. This information is proprietary to Abcam and/or its suppliers.

ポジティブ・コントロール ICC/IF: HepG2 cells.

特記事項 Our RabMAb® technology is a patented hybridoma-based technology for making rabbit monoclonal antibodies. For details on our patents, please refer to **RabMAb**® **patents**.

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outlicensing@thermofisher.com.

#### 製品の特性

製品の状態

保存方法 Shipped at 4°C. Store at +4°C short term (1-2 weeks). Upon delivery aliquot. Store at -20°C.

Avoid freeze / thaw cycle. Store In the Dark.

**バッファー** pH: 7.40

Preservative: 0.02% Sodium azide

Constituents: PBS, 30% Glycerol (glycerin, glycerine), 1% BSA

精製度 Protein A purified

**ポリ/モノ** モノクローナル

**クローン名** E398L **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ab201349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検討ください。

アプリケーション	Abreviews	特記事項
ICC/IF		1/100. This product gave a positive signal in HepG2 cells fixed with 100% methanol (5 min).

#### ターゲット情報

#### 機能

#### 関連疾患

Anchors the lamina and the heterochromatin to the inner nuclear membrane.

Defects in LBR are a cause of Pelger-Huet anomaly (PHA) [MIM:169400]. PHA is an autosomal dominant inherited abnormality of neutrophils, characterized by reduced nuclear segmentation and an apparently looser chromatin structure. Heterozygotes show hypolobulated neutrophil nuclei with coarse chromatin. Presumed homozygous individuals have ovoid neutrophil nuclei, as well as varying degrees of developmental delay, epilepsy, and skeletal abnormalities.

Defects in LBR are the cause of hydrops-ectopic calcification-moth-eaten skeletal dysplasia (HEM) [MIM:215140]; also known as Greenberg skeletal dysplasia. HEM is a rare autosomal recessive chondrodystrophy characterized by early in utero lethality and, therefore, considered to be nonviable. Affected fetuses typically present with fetal hydrops, short-limbed dwarfism, and a marked disorganization of chondro-osseous calcification and may present with polydactyly and additional nonskeletal malformations.

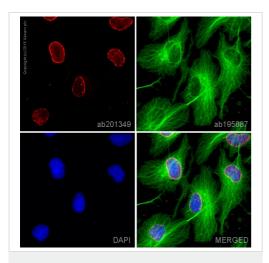
Defects in LBR may be a cause of Reynolds syndrome (REYNS) [MIM:613471]. It is a syndrome specifically associating limited cutaneous systemic sclerosis and primary biliray cirrhosis. It is characterized by liver disease, telangiectasia, abrupt onset of digital paleness or cyanosis in response to cold exposure or stress (Raynaud phenomenon), and variable features of scleroderma. The liver disease is characterized by pruritis, jaundice, hepatomegaly, increased serum alkaline phosphatase and positive serum mitochondrial autoantibodies, all consistent with primary biliary cirrhosis.

## **配列類似性** Belongs to the ERG4/ERG24 family.

#### 翻訳後修飾

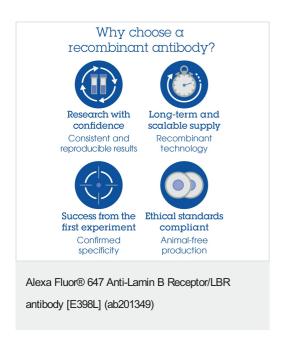
Phosphorylated by CDK1 protein kinase in mitosis when the inner nuclear membrane breaks down into vesicles that dissociate from the lamina and the chromatin. It is phosphorylated by different protein kinases in interphase when the membrane is associated with these structures. Phosphorylation of LBR and HP1 proteins may be responsible for some of the alterations in chromatin organization and nuclear structure which occur at various times during the cell cycle.

#### 画像



Immunocytochemistry/ Immunofluorescence - Alexa Fluor® 647 Anti-Lamin B Receptor/LBR antibody [E398L] (ab201349) ab201349 staining Lamin B Receptor/LBR in HepG2 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 ab201349 at a 1/100 dilution (shown in red) and ab195887, Mouse monoclonal to alpha Tubulin (Alexa Fluor® 488), at a 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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