

Recombinant Human Lamin B Receptor/LBR protein ab132387

画像数 1

製品の詳細

製品名	Recombinant Human Lamin B Receptor/LBR protein
発現系	Wheat germ
アクセッション番号	<u>Q14739</u>
タンパク質長	Full length protein
Animal free	No
由来	Recombinant
生物種	Human
配列	MPSRKFADGEVVRGRWPGSSLYYEVEILSHDSTSQLYTVKYK DGTELELK ENDIKPLTSFRQRKGGSTSSSPSRRRGSRSRSRSPGRPPK SARRSASA SHQADIKEARREVEVKLTPLILKPFGNSISRYNGEPEHIERN DAPHKNTQ EKFSLSQESSYIATQYSLRPRREEVKLKEIDSKEEKYVAKEL AVRTFEVT PIRAKDLEFGGVPGVFLIMFGLPVFLFLLLMCKQKDPSLLN FPPPLPAL YELWETRVFGVYLLWFLIQVLFYLLPIGKVVEGTPLIDGRRL KYRLNGFY AFILTSAVIGTSLFQGVEFHVYSHFLQFALAATVFCVLSV YLYMRSLK APRNDLSPASSGNAVYDFFIGRELNPRIGTFDLKYFCELRPG LIGWVVIN LVMLLAEMKIQDRAVPSLAMILVNSFQLLYVVDALWNEEALL TTMDIIHD GFGFMLAFGDLVWVPFIYSFQAFYLVSHPNESWPMASLIIV LKLCGYVI FRGANSQKNAFRKNPSDPKLAHLKTIHTSTGKNLLVSGWWGF VRHPNYLG DLIMALAWSLPCGFNHILPYFYIIYFTMLLVHREARDEYHCK KKYGVawe KYCQRPYRIFPYIY
予測される分子量	97 kDa including tags
領域	1 to 615

特性

Our **Abpromise guarantee** covers the use of **ab132387** in the following tested applications.

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

アプリケーション

SDS-PAGE

ELISA

Western blot

製品の状態

Liquid

備考

This product was previously labelled as Lamin B Receptor.

前処理および保存

保存方法および安定性

Shipped on dry ice. Upon delivery aliquot and store at -80°C. Avoid freeze / thaw cycles.

pH: 8.00

Constituents: 0.31% Glutathione, 0.79% Tris HCl

関連情報

機能

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

関連疾患

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 biliary 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

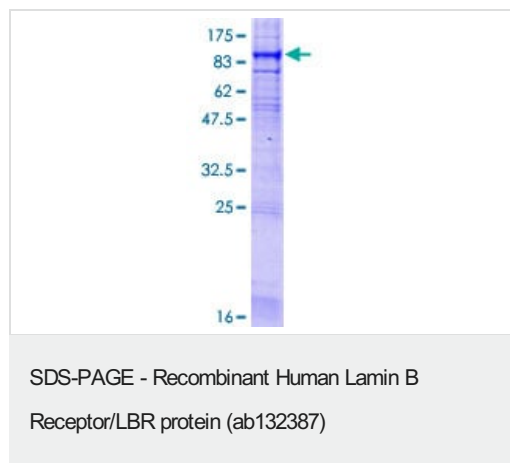
Belongs to the ERG4/ERG24 family.

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.

細胞内局在

Nucleus inner membrane.



ab132387 on 12.5% SDS-PAGE stained with Coomassie Blue.

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

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