

Recombinant Human Lamin B Receptor/LBR protein  
ab132387

1 Image

Description	
Product name	Recombinant Human Lamin B Receptor/LBR protein
Expression system	Wheat germ
Accession	<u>Q14739</u>
Protein length	Full length protein
Animal free	No
Nature	Recombinant
Species	Human
Sequence	MPSRKFADGEVVRGRWPGSSLYEVEILSHDSTSQLYTV KYKDGTELELK ENDIKPLTSFRQQRKGGSTSSSPSRRRGSRSRSRSPGR PPKSARRSASA SHQADIKEARREVEVKLTPLILKPFGNSISRYNGEPEHIERN DAPHKNTQ EKFSLSQESSYIATQYSLRPRREEVKLKEIDSKEEKYVAKE LAVRTFEVT PIRAKDLEFGGVPGVFLIMFGLPVFLFLLLLMCKQKDPSLL NFPPPLPAL YELWETRVFGVYLLWFLIQVLFYLLPIGKVVEGTPLIDGRRL KYRLNGFY AFILTSAVIGTSLFQGVEFHVYSHFLQFALAATVFCVVLVS YLYMRSLK APRNDLSPASSGNAVYDFFIGRELNPRIGTFDLKYFCELRP GLIGWVVIN LVMLLAEMKIQDRAVPSLAMILVNSFQLLYVVDALWNEEA LLTTMDIHD GFGFMLAFGDLVWVPFIYSFQAFYLVSHPNEVSWPMASLI MLKLCGYVI FRGANSQKNAFRKNPSDPKLAHLKTIHTSTGKNLLVSGW WGFVRHPNYLG DLIMALAWSLPCGFNHILPYFYIYFTMLLVHREARDEYHCK KKYGVawe KYCQRVPYRIFPIY
Predicted molecular weight	97 kDa including tags

## Specifications

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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.

<b>Applications</b>	SDS-PAGE
	ELISA
	Western blot
<b>Form</b>	Liquid
<b>Additional notes</b>	This product was previously labelled as Lamin B Receptor.

## Preparation and Storage

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<b>Stability and Storage</b>	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

## General Info

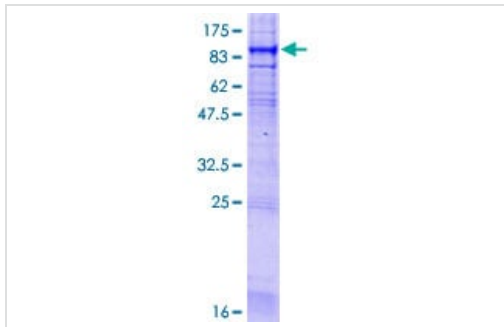
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<b>Function</b>	Anchors the lamina and the heterochromatin to the inner nuclear membrane.
<b>Involvement in disease</b>	<p>Defects in LBR are a cause of Pelger-Huet anomaly (PHA) [MIM:169400]. PHA is an autosomal dominant inherited abnormality of neutrophils, 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.</p> <p>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.</p> <p>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.</p>
<b>Sequence similarities</b>	Belongs to the ERG4/ERG24 family.
<b>Post-translational modifications</b>	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.

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## Images

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

SDS-PAGE - Recombinant Human Lamin B  
Receptor/LBR protein (ab132387)

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

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## Our Abpromise to you: Quality guaranteed and expert technical support

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- Replacement or refund for products not performing as stated on the datasheet
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If the product does not perform as described on this datasheet, we will offer a refund or replacement. For full details of the Abpromise, please visit <https://www.abcam.com/abpromise> or contact our technical team.

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