

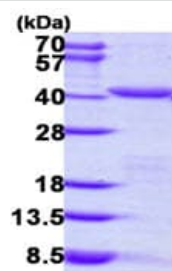
Recombinant Human PEX19 protein ab111623

1 Image

Description	
Product name	Recombinant Human PEX19 protein
Purity	> 90 % SDS-PAGE. ab111623 is purified using conventional chromatography techniques.
Expression system	Escherichia coli
Accession	<b><u>P40855</u></b>
Protein length	Full length protein
Animal free	No
Nature	Recombinant
Species	Human
Sequence	<b>MGSSHHHHHHSSGLVPRGSHMAAAEEGCSVGAEADRE</b> <b>LEELLESALDDFD</b> <b>KAKPSPAPPSTTTAPDASGPQKRSPGDTAKDALFASQEK</b> <b>FFQELFDSELA</b> <b>SQATAEFEKAMKELAEEEPHLVEQFQKLSEAAGRVGSD</b> <b>MTSQQEFTSCLK</b> <b>ETLSGLAKNATDLQNSSMSEEELTKAMEGLGMDEGDGE</b> <b>GNILPIMQSIMQ</b> <b>NLLSKDVLVPSLKEITEKYPEWLQSHRESLPPEQFEKYQE</b> <b>QHSVMCKICE</b> <b>QFEAETPTDSETTQKARFEMVLDLMQQLQDLGHPPKELA</b> <b>GEMPPGLNFDL DALNLSGPPGASGEQC</b>
Predicted molecular weight	35 kDa including tags
Amino acids	1 to 296
Tags	His tag N-Terminus

Specifications	
Our <b><u>Abpromise guarantee</u></b> covers the use of <b>ab111623</b> in the following tested applications.	
The application notes include recommended starting dilutions; optimal dilutions/concentrations should be determined by the end user.	
Applications	SDS-PAGE  Mass Spectrometry
Mass spectrometry	MALDI-TOF

<b>Form</b>	Liquid
<b>Preparation and Storage</b>	
<b>Stability and Storage</b>	<p>Shipped at 4°C. Upon delivery aliquot and store at -20°C or -80°C. Avoid repeated freeze / thaw cycles.</p> <p>pH: 8.00</p> <p>Constituents: 0.32% Tris HCl, 10% Glycerol (glycerin, glycerine)</p>
<b>General Info</b>	
<b>Function</b>	Necessary for early peroxisomal biogenesis. Acts both as a cytosolic chaperone and as an import receptor for peroxisomal membrane proteins (PMPs). Binds and stabilizes newly synthesized PMPs in the cytoplasm by interacting with their hydrophobic membrane-spanning domains, and targets them to the peroxisome membrane by binding to the integral membrane protein PEX3. Excludes CDKN2A from the nucleus and prevents its interaction with MDM2, which results in active degradation of TP53.
<b>Tissue specificity</b>	Ubiquitously expressed. Isoform 1 is strongly predominant in all tissues except in utero where isoform 2 is the main form.
<b>Involvement in disease</b>	<p>Defects in PEX19 are the cause of peroxisome biogenesis disorder complementation group 14 (PBD-CG14) [MIM:600279]; also known as PBD-CGJ. PBD refers to a group of peroxisomal disorders arising from a failure of protein import into the peroxisomal membrane or matrix. The PBD group is comprised of four disorders: Zellweger syndrome (ZWS), neonatal adrenoleukodystrophy (NALD), infantile Refsum disease (IRD), and classical rhizomelic chondrodysplasia punctata (RCDP). ZWS, NALD and IRD are distinct from RCDP and constitute a clinical continuum of overlapping phenotypes known as the Zellweger spectrum. The PBD group is genetically heterogeneous with at least 14 distinct genetic groups as concluded from complementation studies.</p> <p>Defects in PEX19 are a cause of Zellweger syndrome (ZWS) [MIM:214100]. ZWS is a fatal peroxisome biogenesis disorder characterized by dysmorphic facial features, hepatomegaly, ocular abnormalities, renal cysts, hearing impairment, profound psychomotor retardation, severe hypotonia and neonatal seizures. Death occurs within the first year of life.</p>
<b>Sequence similarities</b>	Belongs to the peroxin-19 family.
<b>Cellular localization</b>	Cytoplasm. Peroxisome membrane. Mainly cytoplasmic. Some fraction membrane-associated to the outer surface of peroxisomes.
<b>Images</b>	



SDS-PAGE - Recombinant Human PEX19 protein  
(ab111623)

15% SDS-PAGE showing ab111623 at approximately 34.6kDa  
(3µg).  
(Molecular weight on SDS-PAGE will appear higher)

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

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

---

- Replacement or refund for products not performing as stated on the datasheet
- Valid for 12 months from date of delivery
- Response to your inquiry within 24 hours
- We provide support in Chinese, English, French, German, Japanese and Spanish
- Extensive multi-media technical resources to help you
- We investigate all quality concerns to ensure our products perform to the highest standards

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.

### Terms and conditions

---

- Guarantee only valid for products bought direct from Abcam or one of our authorized distributors