abcam

Product datasheet

Recombinant Human PEX19 protein ab111623

1 图像

描述

产品名称	重组人PEX19蛋白
纯度	> 90 % SDS-PAGE. ab111623 is purified using conventional chromatography techniques.
表达系统	Escherichia coli
Accession	P40855
蛋白长度	Full length protein
无动物成分	No
性质	Recombinant
种属	Human
序列	MGSSHHHHHHSSGLVPRGSHMAAAEEGCSVGAEADRELEE LLESALDDFD KAKPSPAPPSTTTAPDASGPQKRSPGDTAKDALFASQEKFFQ ELFDSELA SQATAEFEKAMKELAEEEPHLVEQFQKLSEAAGRVGSDMTSQ QEFTSCLK ETLSGLAKNATDLQNSSMSEEELTKAMEGLGMDEGDGEGNIL PIMQSIMQ NLLSKDVLYPSLKEITEKYPEWLQSHRESLPPEQFEKYQEQH SVMCKICE QFEAETPTDSETTQKARFEMVLDLMQQLQDLGHPPKELAGEM PPGLNFDL DALNLSGPPGASGEQC
预测分子量	35 kDa including tags
氨基酸	1 to 296
标签	His tag N-Terminus

技术指标

Our **Abpromise guarantee** covers the use of **ab111623** in the following tested applications.

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

应用	SDS-PAGE
	Mass Spectrometry
质谱法	MALDI-TOF

形式	Liquid

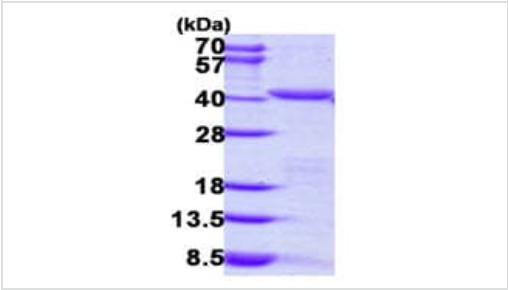
制备和贮存

稳定性和存储	Shipped at 4°C. Upon delivery aliquot and store at -20°C or -80°C. Avoid repeated freeze / thaw cycles. pH: 8.00 Constituents: 0.32% Tris HCl, 10% Glycerol (glycerin, glycerine)

常规信息

功能	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.
组织特异性	Ubiquitously expressed. Isoform 1 is strongly predominant in all tissues except in utero where isoform 2 is the main form.
疾病相关	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. 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.
序列相似性	Belongs to the peroxin-19 family.
细胞定位	Cytoplasm. Peroxisome membrane. Mainly cytoplasmic. Some fraction membrane-associated to the outer surface of peroxisomes.

图片



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)

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.cn/abpromise** or contact our technical team.

Terms and conditions

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