abcam

Product datasheet

Recombinant Human PEX19 protein ab111623

画像数 1

製品の詳細

製品名	Recombinant Human PEX19 protein
精製度	> 90 % SDS-PAGE. ab111623 is purified using conventional chromatography techniques.
発現系	Escherichia coli
アクセッション番号	P40855
タンパク質長	Full length protein
Animal free	No
由来	Recombinant
生物種	Human
配列	MGSSHHHHHHSSGLVPRGSHMAAAEEGCSVGAEADRELEE LLESALDDFD KAKPSPAPPSTTTAPDASGPQKRSPGDTAKDALFASQEKFFQ ELFDSELA SQATAEFEKAMKELAEEEPHLVEQFQKLSEAAGRVGSDMTSQ QEFTSCLK ETLSGLAKNATDLQNSSMSEEELTKAMEGLGMDEGDGEGNIL PIMQSIMQ NLLSKDVLYPSLKEITEKYPEWLQSHRESLPPEQFEKYQEQH SVMCKICE QFEAETPTDSETTQKARFEMVLDLMQQLQDLGHPPKELAGEM PPGLNFDL DALNLSGPPGASGEQC
予測される分子量	35 kDa including tags
領域	1 to 296
タグ	His tag N-Terminus

特性

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.

アプリケーション	SDS-PAGE Mass Spectrometry
質量分析	MALDI-TOF

製品の状態	Liquid

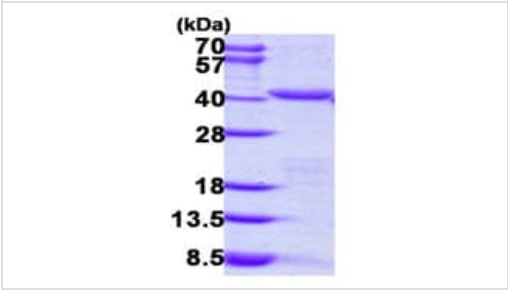
前処理および保存

保存方法および安定性	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)

関連情報

機能	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.
組織特異性	Ubiquitously expressed. Isoform 1 is strongly predominant in all tissues except in utero where isoform 2 is the main form.
関連疾患	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.
配列類似性	Belongs to the peroxin-19 family.
細胞内局在	Cytoplasm. Peroxisome membrane. Mainly cytoplasmic. Some fraction membrane-associated to the outer surface of peroxisomes.

画像



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)

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