abcam

Product datasheet

Anti-PEX19 antibody ab116358

画像数 1

製品の概要

製品名	Anti-PEX19 antibody
製品の詳細	Rabbit polyclonal to PEX19
由来種	Rabbit
アプリケーション	**適用あり:** WB
種交差性	**交差種:** Human **交差が予測される動物種:** Mouse, Rat, Rabbit, Horse, Guinea pig, Cow, Cat, Dog, Pig
免疫原	Synthetic peptide corresponding to a region within C terminal amino acids 234-283 (AETPTDSETT QKARFEMVLD LMQQLQDLGH PPKELAGEMP PGLNFDLDAL) of Human PEX19 (NP_002848). Run BLAST with ExPASy Run BLAST with NCBI
ポジティブ・コントロール	HCT15 cell lysate.

製品の特性

製品の状態	Liquid
保存方法	Shipped at 4°C. Upon delivery aliquot and store at -20°C. Avoid repeated freeze / thaw cycles.
バッファー	Preservative: 0.09% Sodium azide Constituents: 2% Sucrose, PBS
精製度	Immunogen affinity purified
ポリ/モノ	ポリクローナル
アイソタイプ	IgG

アプリケーション

Our Abpromise guarantee covers the use of **ab116358** 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	Abreviews	特記事項

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WB		Use a concentration of 1 µg/ml. Predicted molecular weight: 33 kDa. Good results were obtained when blocked with 5% non-fat dry milk in 0.05% PBS-T.

ターゲット情報

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.

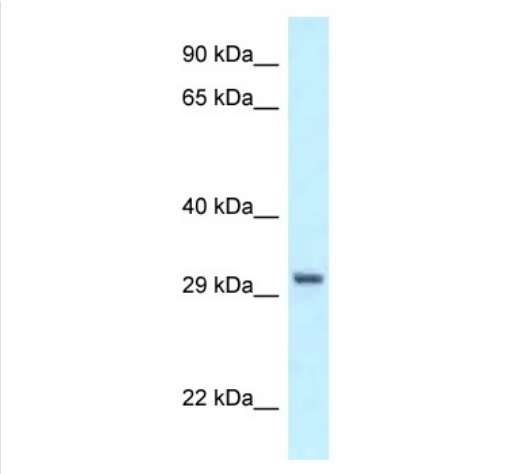
配列類似性

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



Western blot - Anti-PEX19 antibody (ab116358)

Anti-PEX19 antibody (ab116358) at 1 µg/ml + HCT15 cell lysate at 10 µg

Predicted band size: 33 kDa

Gel concentration: 12%.

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