abcam

Product datasheet

Anti-PEX14 antibody ab76862

2 图像

概述

产品名称	Anti-PEX14抗体
描述	**小鼠单克隆抗体to PEX14**
宿主	Mouse
经测试应用	**适用于:** WB, ELISA
种属反应性	**与反应:** Recombinant fragment **预测可用于:** Human
免疫原	Recombinant fragment: LGPQEEGEGV VDVKGQVRME VQGEEEKRED KEDEEDEEDD DVSHVDEEDC LGVQREDRRG GDGQINEQVE KLRRPEGASN ESE, corresponding to amino acids 293-375 of human PEX14 (NP_004556) with a 26 kDa tag. Run BLAST with ExPASy Run BLAST with NCBI

性能

形式	Liquid
存放说明	Shipped at 4°C. Upon delivery aliquot and store at -20°C or -80°C. Avoid repeated freeze / thaw cycles.
存储溶液	Preservative: None Constituents: 1X PBS, pH 7.2
纯度	Protein A purified
克隆	单克隆
同种型	IgG2a
轻链类型	kappa

应用

Our Abpromise guarantee covers the use of **ab76862** in the following tested applications.

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

应用	Ab 评论	说明
WB		Use a concentration of 1 - 5 µg/ml. Predicted molecular weight: 41 kDa. This antibody has only been tested in WB against the recombinant fragment used as immunogen. We have no data on the detection of endogenous protein.
ELISA		Use at an assay dependent dilution. Detection limit ~3ng/ml when used as a capture antibody.

靶标

功能

Component of the peroxisomal translocation machinery with PEX13 and PEX17. Interacts with both the PTS1 and PTS2 receptors. Binds directly to PEX17.

疾病相关

Defects in PEX14 are the cause of peroxisome biogenesis disorder complementation group K (PBD-CGK) [MIM:601791]. PBD-CGK is a peroxisomal disorder arising from a failure of protein import into the peroxisomal membrane or matrix. The peroxisome biogenesis disorders (PBD group) are genetically heterogeneous with at least 14 distinct genetic groups as concluded from complementation studies. Include disorders are: 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 (PBD-ZSS).
Defects in PEX14 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.

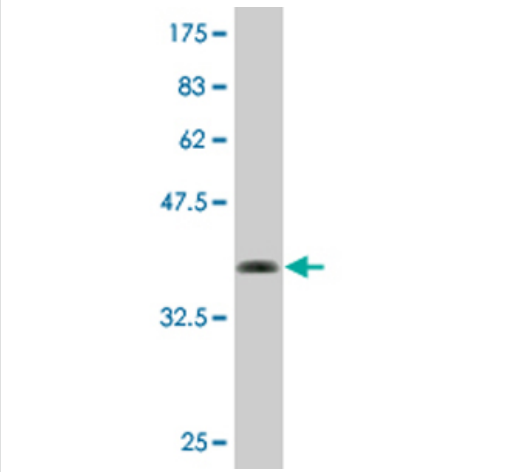
序列相似性

Belongs to the peroxin-14 family.

细胞定位

Peroxisome membrane.

图片

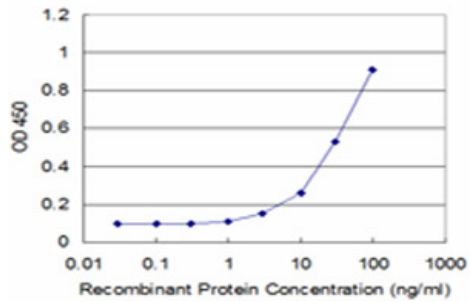


Western blot - Anti-PEX14 antibody (ab76862)

Anti-PEX14 antibody (ab76862) at 5 µg/ml + immunogen at 0.2 µg

Secondary
Goat anti-mouse IgG (H&L)-HRP conjugate at 1/5000 dilution

Predicted band size: 41 kDa
Observed band size: 35 kDa



ELISA - Anti-PEX14 antibody (ab76862)

Detection limit for ab76862 is approximately 3ng/ml as a capture antibody.

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