abcam

Product datasheet

Anti-PAF-1 antibody ab95289

1 图像

概述

产品名称	Anti-PAF-1抗体
描述	兔多克隆抗体to PAF-1
宿主	Rabbit
经测试应用	**适用于:** WB
种属反应性	**与反应:** Mouse, Human
免疫原	KLH conjugated synthetic peptide selected from the central region of Human PEX2 (NP_000309.1)
阳性对照	Mouse cerebellum tissue lysates

性能

形式	Liquid
存放说明	Shipped at 4°C. Upon delivery aliquot and store at -20°C. Avoid repeated freeze / thaw cycles.
存储溶液	Preservative: 0.09% Sodium azide Constituent: PBS
纯度	Immunogen affinity purified
纯化说明	This antibody is purified through a protein A column, followed by peptide affinity purification.
克隆	多克隆
同种型	IgG

应用

The Abpromise guarantee **Abpromise™承诺保证使用ab95289于以下的经测试应用**

"应用说明"部分 下显示的仅为推荐的起始稀释度;实际最佳的稀释度/浓度应由使用者检定。

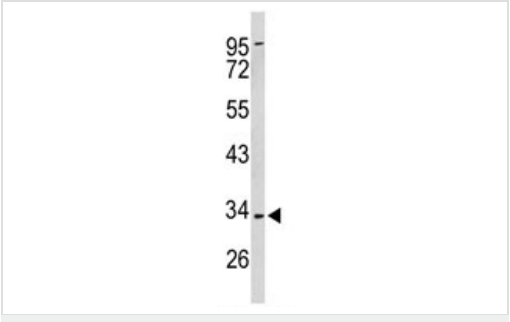
应用	Ab评论	说明
WB		1/100 - 1/500. Predicted molecular weight: 35 kDa.

靶标

靶标

功能	Somewhat implicated in the biogenesis of peroxisomes.
疾病相关	Defects in PEX2 are the cause of peroxisome biogenesis disorder complementation group 5 (PBD-CG5) [MIM:170993]; also known as PBD-CGF. 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 PEX2 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. Defects in PEX2 are a cause of infantile Refsum disease (IRD) [MIM:266510]. IRD is a mild peroxisome biogenesis disorder (PBD). Clinical features include early onset, mental retardation, minor facial dysmorphism, retinopathy, sensorineural hearing deficit, hepatomegaly, osteoporosis, failure to thrive, and hypocholesterolemia. The biochemical abnormalities include accumulation of phytanic acid, very long chain fatty acids (VLCFA), di- and trihydroxycholestanoic acid and pipecolic acid.
序列相似性	Belongs to the pex2/pex10/pex12 family. Contains 1 RING-type zinc finger.
细胞定位	Peroxisome membrane.

图片



Western blot - Anti-PEX2 antibody (ab95289)

Anti-PAF-1 antibody (ab95289) at 1/100 dilution + Mouse cerebellum tissue lysate at 35 µg

Predicted band size: 35 kDa

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