产品名称: Peroxin 1 Rabbit Polyclonal Antibody

产品货号: APRab15980



#### 产品概述 (Summary)

产品名称 (Production Name) Peroxin 1 Rabbit Polyclonal Antibody

描述 (Description) Rabbit polyclonal Antibody

宿主 (Host) Rabbit

应用 (Application)IHC,ICC/IF,ELISA种属反应性 (Reactivity)Human,Mouse

### 产品性能 (Performance)

偶联物 (Conjugation)Unconjugated修饰 (Modification)Unmodified

同种型 (Isotype) IgG

克隆 (Clonality) Polyclonal 形式 (Form) Liquid

Store at 4°C short term. Aliquot and store at -20°C long term. Avoid 存放说明 (Storage)

freeze/thaw cycles.

Liquid in PBS containing 50% glycerol, 0.5% protective protein and 0.02% 储存溶液 (Buffer)

New type preservative N.

纯化方式 (Purification) Affinity purification

# 免疫原信息 (Immunogen)

基因名 (Gene Name) PEX1

PEX1; Peroxisome biogenesis factor 1; Peroxin-1; Peroxisome biogenesis **别名 (Alternative Names)** 

disorder protein 1

基因 ID (Gene ID) 5189.0

O43933.The antiserum was produced against synthesized peptide derived 蛋白ID (SwissProt ID)

from human PEX1. AA range:1234-1283

# 产品应用 (Application)

稀释比 (Dilution Ratio) IHC 1:100-1:300,ICC/IF 1:50-1:200,ELISA 1:5000-1:20000

蛋白分子量(Molecular Weight)

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#### 研究背景 (Background)

This gene encodes a member of the AAA ATPase family, a large group of ATPases associated with diverse cellular activities. This protein is cytoplasmic but is often anchored to a peroxisomal membrane where it forms a heteromeric complex and plays a role in the import of proteins into peroxisomes and peroxisome biogenesis. Mutations in this gene have been associated with complementation group 1 peroxisomal disorders such as neonatal adrenoleukodystrophy, infantile Refsum disease, and Zellweger syndrome. Alternatively spliced transcript variants have been found for this gene. [provided by RefSeq, Sep 2013], disease: Defects in PEX1 are a cause of adrenoleukodystrophy neonatal (NALD) [MIM:202370]. NALD is a peroxisome biogenesis disorder (PBD) characterized by the accumulation of very long-chain fatty acids, adrenal insufficiency and mental retardation, disease: Defects in PEX1 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., disease: Defects in PEX1 are the cause of peroxisome biogenesis disorder complementation group 1 (PBD-CG1) [MIM:602136]; also known as PBD-CGE. 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, function: Required for stability of PEX5 and protein import into the peroxisome matrix. Anchored by PEX26 to peroxisome membranes, possibly to form heteromeric AAA ATPase complexes required for the import of proteins into peroxisomes, PTM: Phosphorylated upon DNA damage, probably by ATM or ATR., similarity: Belongs to the AAA ATPase family., subcellular location: Associated with peroxisomal membranes., subunit: Interacts directly with PEX6. Interacts indirectly with PEX26, via its interaction with PEX6.,

#### 研究领域(Research Area)

Signal Transduction; Protein Trafficking; Organelle Proteins

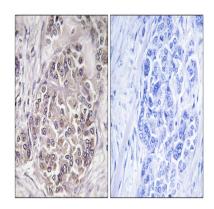
图片 (Image Data)

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Immunohistochemistry analysis of paraffin-embedded human breast carcinoma tissue, using PEX1 Antibody. The picture on the right is blocked with the synthesized peptide.

### 注意事项 (Note)

For research use only.

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