产品名称: PEX12 Rabbit Polyclonal Antibody

产品货号: APRab15998



# 产品概述 (Summary)

产品名称 (Production Name) PEX12 Rabbit Polyclonal Antibody

描述 (Description) Rabbit polyclonal Antibody

宿主 (Host) Rabbit 应用 (Application) WB,ELISA

种属反应性 (Reactivity) Human, Rat, 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.

储存溶液 (Buffer) Liquid in PBS containing 50% glycerol, and 0.02% New type preservative N.

**纯化方式 (Purification)** Affinity purification

# 免疫原信息 (Immunogen)

基因名 (Gene Name) PEX12 PAF3

别名 (Alternative Names)

基因 ID (Gene ID) 5193.0

O00623.Synthesized peptide derived from human protein . at AA range: 180-蛋白ID (SwissProt ID)

260

### 产品应用(Application)

**稀释比 (Dilution Ratio)** WB 1:500-1:2000,ELISA 1:5000-1:20000

蛋白分子量 (Molecular Weight) 39kDa

## 研究背景 (Background)

peroxisomal biogenesis factor 12(PEX12) Homo sapiens This gene belongs to the peroxin-12 family. Peroxins (PEXs) are

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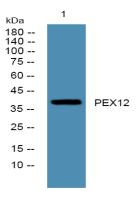
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proteins that are essential for the assembly of functional peroxisomes. The peroxisome biogenesis disorders (PBDs) are a group of genetically heterogeneous autosomal recessive, lethal diseases characterized by multiple defects in peroxisome function. The peroxisomal biogenesis disorders are a heterogeneous group with at least 14 complementation groups and with more than 1 phenotype being observed in cases falling into particular complementation groups. Although the clinical features of PBD patients vary, cells from all PBD patients exhibit a defect in the import of one or more classes of peroxisomal matrix proteins into the organelle. Defects in this gene are a cause of Zellweger syndrome (ZWS). [provided by RefSeq, Oct 2008], disease: Defects in PEX12 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., disease: Defects in PEX12 are the cause of peroxisome biogenesis disorder complementation group 3 (PBD-CG3) [MIM:601758]. 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 protein import into peroxisomes, similarity: Belongs to the pex2/pex10/pex12 family, similarity: Contains 1 RING-type zinc finger, subunit: Interacts with PEX5 and PEX10. Interacts with PEX19 via its cytoplasmic domain.,

### 研究领域 (Research Area)

#### 图片 (Image Data)



Western blot analysis of lysates from KB cells, primary antibody was diluted at 1:1000, 4° over night

### 注意事项 (Note)

For research use only.

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