

产品名称: Peroxin 5 Rabbit Polyclonal Antibody
产品货号: AP Rab15988



产品概述 (Summary)

产品名称 (Production Name)	Peroxin 5 Rabbit Polyclonal Antibody
描述 (Description)	Rabbit polyclonal Antibody
宿主 (Host)	Rabbit
应用 (Application)	WB,IHC
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
存放说明 (Storage)	Store at 4°C short term. Aliquot and store at -20°C long term. Avoid freeze/thaw cycles.
储存溶液 (Buffer)	Liquid in PBS containing 50% glycerol, 0.5% protective protein 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)	PEX5 PEX5; PXR1; Peroxisomal targeting signal 1 receptor; PTS1 receptor; PTS1R;
别名 (Alternative Names)	PTS1-BP; Peroxin-5; Peroxisomal C-terminal targeting signal import receptor; Peroxisome receptor 1
基因 ID (Gene ID)	5830.0
蛋白 ID (SwissProt ID)	P50542.Synthesized peptide derived from Peroxin 5 . at AA range: 540-620

产品应用 (Application)

稀释比 (Dilution Ratio)	WB 1:500-1:2000,IHC 1:50-1:300
蛋白分子量 (Molecular Weight)	70kDa

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研究背景 (Background)

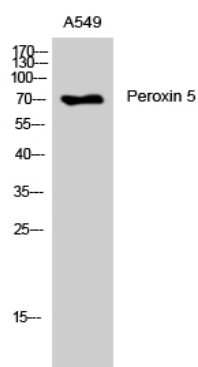
The product of this gene binds to the C-terminal PTS1-type tripeptide peroxisomal targeting signal (SKL-type) and plays an essential role in peroxisomal protein import. Peroxisins (PEXs) are proteins that are essential for the assembly of functional peroxisomes. The peroxisome biogenesis disorders (PBDs) 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 neonatal adrenoleukodystrophy (NALD) disease: Defects in PEX5 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. Inheritance is autosomal recessive. disease: Defects in PEX5 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 PEX5 may be 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 pipelicolic acid. function: Binds to the C-terminal PTS1-type tripeptide peroxisomal targeting signal (SKL-type) and plays an essential role in peroxisomal protein import. similarity: Belongs to the peroxisomal targeting signal receptor family. similarity: Contains 7 TPR repeats. subcellular location: Its distribution appears to be dynamic. It is probably a cycling receptor found mainly in the cytoplasm and as well associated to the peroxisomal membrane through a docking factor (PEX13). subunit: Interacts with PEX7 and PEX13 (By similarity). Interacts with PEX12 and PEX14. tissue specificity: Detected in heart, brain, placenta, lung, liver, skeletal muscle, kidney and pancreas.

研究领域 (Research Area)

Signal Transduction; Protein Trafficking; Organelle Proteins

图片 (Image Data)

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Western Blot analysis of A549 cells using Peroxin 5 Polyclonal Antibody

注意事项 (Note)

For research use only .