Product Name: Peroxin 5 Rabbit Polyclonal Antibody
Catalog #: APRab15988

Summary

Production Name	Peroxin 5 Rabbit Polyclonal Antibody
Description	Rabbit Polyclonal Antibody
Host	Rabbit
Application	WB
Reactivity	Human,Mouse

Performance

Conjugation	Unconjugated
Modification	Unmodified
Isotype	IgG
Clonality	Polyclonal
Form	Liquid
Storage	Store at 4°C short term. Aliquot and store at -20°C long term. Avoid freeze/thaw cycles.
Buffer	Liquid in PBS containing 50% glycerol, 0.5% BSA and 0.02% New type preservative N.
Purification	Affinity purification

Immunogen

Gene Name	PEX5
Alternative Names	PEX5; PXR1; Peroxisomal targeting signal 1 receptor; PTS1 receptor; PTS1R; PTS1-BP; Peroxin-5; Peroxisomal C-terminal targeting signal import receptor; Peroxisome receptor 1
Gene ID	5830.0
SwissProt ID	P50542.Synthesized peptide derived from Peroxin 5 . at AA range: 540-620

Application

Dilution Ratio	WB 1:500-2000
Molecular Weight	70kD

Background

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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. Peroxins (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 pipecolic 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.,

Research Area

Image Data

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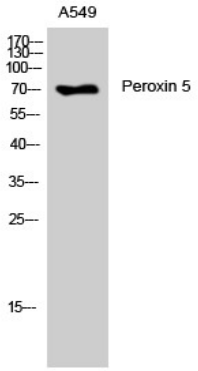

Western Blot analysis of A549 cells using Peroxin 5 Polyclonal Antibody

Note

For research use only.