

Product Name: Peroxin 1 Rabbit Polyclonal Antibody
Catalog #: APRab15980



Summary

Production Name	Peroxin 1 Rabbit Polyclonal Antibody
Description	Rabbit Polyclonal Antibody
Host	Rabbit
Application	IHC,ELISA
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	PEX1
Alternative Names	PEX1; Peroxisome biogenesis factor 1; Peroxin-1; Peroxisome biogenesis disorder protein 1
Gene ID	5189.0
SwissProt ID	O43933.The antiserum was produced against synthesized peptide derived from human PEX1. AA range:1234-1283

Application

Dilution Ratio	IHC 1:100-1:300 ELISA: 1:10000
Molecular Weight	

Background

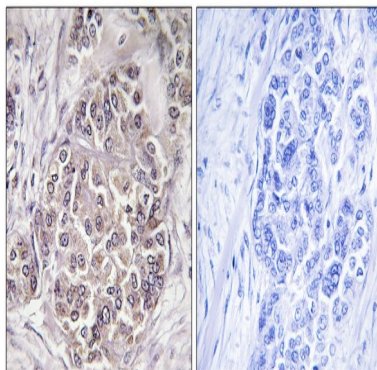
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This gene encodes a member of the AAA ATPase family, a large group of ATPases associated with diverse cellular activities. This protein is cytoplasmic but is often anchored to a peroxisomal membrane where it forms a heteromeric complex and plays a 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 pipercolic 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.,

Research Area

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.

Note

For research use only.