10320 Camino Santa Fe, Suite G
San Diego, CA 92121
Tel: 858.875.1900 Fax: 858.875.1999

PEX13 Rabbit pAb

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Catalog # AP54905

Product Information

Application	IHC-P, IHC-F, IF
Primary Accession	Q92968
Reactivity	Rat, Mouse
Predicted	Dog, Human, Rabbit, Sheep
Host	Rabbit
Clonality	Polyclonal
Calculated MW	44130
Physical State	Liquid
Immunogen	KLH conjugated synthetic peptide derived from human PEX13
Epitope Specificity	251-350/403
Isotype	IgG
Purity	affinity purified by Protein A
Buffer	0.01M TBS (pH7.4) with 1% BSA, 0.02% Proclin300 and 50% Glycerol.
SUBCELLULAR LOCATION	Peroxisome membrane.
DISEASE	Defects in PEX13 are the cause of peroxisome biogenesis disorder complementation group 13 (PBD-CG13) [MIM:601789]; also known as PBD-CGH. PBD-CG13 is a peroxisomal disorder arising from a failure of protein import into the peroxisomal membrane or matrix. The peroxisome biogenesis disorders (PBD group) are genetically heterogeneous with at least 14 distinct genetic groups as concluded from complementation studies. Include disorders are: 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 (PBD-ZSS). Defects in PEX13 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.
Important Note	This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.
Background Descriptions	This gene encodes a peroxisomal membrane protein that binds the type 1 peroxisomal targeting signal receptor via a SH3 domain located in the cytoplasm. Mutations and deficiencies in peroxisomal protein importing and peroxisome assembly lead to peroxisomal biogenesis disorders, an example of which is Zellweger syndrome. [provided by RefSeq, Oct 2008]

Additional Information

Gene ID	5194
Other Names	Peroxisomal membrane protein PEX13, Peroxin-13, PEX13

	{ECO:0000303|PubMed:9653144, ECO:0000312|HGNC:HGNC:8855}
Dilution	IHC-P=1:100-500,IHC-F=1:100-500,IF=1:100-500
Storage	Store at -20 °C for one year. Avoid repeated freeze/thaw cycles. When reconstituted in sterile pH 7.4 0.01M PBS or diluent of antibody the antibody is stable for at least two weeks at 2-4 °C.

Protein Information

Name	PEX13 {ECO:0000303|PubMed:9653144, ECO:0000312|HGNC:HGNC:8855}
Function	Component of the PEX13-PEX14 docking complex, a translocon channel that specifically mediates the import of peroxisomal cargo proteins bound to PEX5 receptor (PubMed:28765278, PubMed:8858165, PubMed:9653144). The PEX13-PEX14 docking complex forms a large import pore which can be opened to a diameter of about 9 nm (By similarity). Mechanistically, PEX5 receptor along with cargo proteins associates with the PEX14 subunit of the PEX13-PEX14 docking complex in the cytosol, leading to the insertion of the receptor into the organelle membrane with the concomitant translocation of the cargo into the peroxisome matrix (PubMed:28765278, PubMed:8858165, PubMed:9653144). Involved in the import of PTS1- and PTS2-type containing proteins (PubMed:8858165, PubMed:9653144).
Cellular Location	Peroxisome membrane; Multi-pass membrane protein

Background

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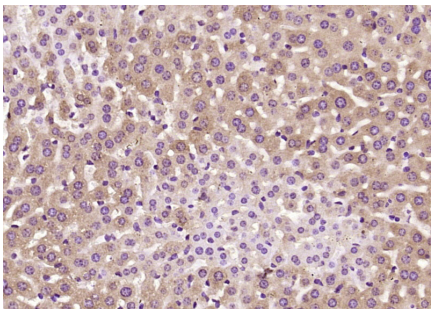
Images



Paraformaldehyde-fixed, paraffin embedded (mouse liver); Antigen retrieval by boiling in sodium citrate buffer (pH6.0) for 15min; Block endogenous peroxidase by 3% hydrogen peroxide for 20 minutes; Blocking buffer (normal goat serum) at 37°C for 30min; Antibody incubation with (PEX13) Polyclonal Antibody, Unconjugated (AP54905) at 1:200 overnight at 4°C, followed by operating according to SP Kit(Rabbit) (sp-0023) instructionsand DAB staining.