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PEX14 Rabbit pAb

Catalog # AP99346

Product Information

Application	IHC-P, IHC-F, IF
Primary Accession	O75381
Other Accession	O75381
Reactivity	Rat
Predicted	Human, Mouse, Rabbit, Chicken, Dog, Horse, Sheep
Host	Rabbit
Clonality	Polyclonal
Calculated MW	41237
Physical State	Liquid
Immunogen	KLH conjugated synthetic peptide derived from human PEX14
Epitope Specificity	151-250/377
Isotype	IgG
Purity	affinity purified by Protein A
Buffer	0.01M TBS (pH7.4) with 1% BSA, 0.02% Proclin300 and 50% Glycerol.
SIMILARITY	Belongs to the peroxin-14 family.
DISEASE	Defects in PEX14 are the cause of peroxisome biogenesis disorder complementation group K (PBD-CGK) [MIM:601791]. PBD-CGK 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 PEX14 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.
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 an essential component of the peroxisomal import machinery. The protein is integrated into peroxisome membranes with its C-terminus exposed to the cytosol, and interacts with the cytosolic receptor for proteins containing a PTS1 peroxisomal targeting signal. The protein also functions as a transcriptional corepressor and interacts with a histone deacetylase. A mutation in this gene results in one form of Zellweger syndrome. [provided by RefSeq, Jul 2008]

Additional Information

Gene ID	5195

Other Names	NAPP2; PBD13A; Pex14p; dJ734G22.2; PEX14_HUMAN; PEX14; PTS1 receptor-docking protein; Peroxin-14; Peroxisomal membrane anchor protein PEX14; PEX14_MOUSE; PEX14_RAT; peroxisomal biogenesis factor 14
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	PEX14 {ECO:0000303|PubMed:9653144, ECO:0000312|HGNC:HGNC:8856}
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:24235149, PubMed:28765278, 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:24235149, PubMed:28765278). Plays a key role for peroxisome movement through a direct interaction with tubulin (PubMed:21525035).
Cellular Location	Peroxisome membrane; Single-pass membrane protein {ECO:0000250|UniProtKB:Q642G4}

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