

PEX6 Rabbit pAb

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Catalog # AP54907

Product Information

Application	IHC-P, IHC-F, IF
Primary Accession	Q13608
Reactivity	Mouse
Predicted	Rat, Pig, Human, Rabbit, Horse
Host	Rabbit
Clonality	Polyclonal
Calculated MW	104061
Physical State	Liquid
Immunogen	KLH conjugated synthetic peptide derived from human PEX6
Epitope Specificity	751-850/980
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	Cytoplasm. Peroxisome membrane. Note: Associated with peroxisomal membranes.
DISEASE	The disease is caused by mutations affecting the gene represented in this entry. Disease description: 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).
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 member of the AAA (ATPases associated with diverse cellular activities) family of ATPases. This member is a predominantly cytoplasmic protein, which plays a direct role in peroxisomal protein import and is required for PTS1 (peroxisomal targeting signal 1, a C-terminal tripeptide of the sequence ser-lys-leu) receptor activity. Mutations in this gene cause peroxisome biogenesis disorders of complementation group 4 and complementation group 6. [provided by RefSeq, Oct 2008]

Additional Information

Gene ID	5190
Other Names	Peroxisomal ATPase PEX6, 3.6.4.-, Peroxin-6, Peroxisomal biogenesis factor 6, Peroxisomal-type ATPase 1, Peroxisome assembly factor 2, PAF-2, PEX6

{ECO:0000303 | PubMed:10408779, ECO:0000312 | HGNC:HGNC:8859}

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

PEX6 {ECO:0000303 | PubMed:10408779, ECO:0000312 | HGNC:HGNC:8859}

Function

Component of the PEX1-PEX6 AAA ATPase complex, a protein dislocase complex that mediates the ATP-dependent extraction of the PEX5 receptor from peroxisomal membranes, an essential step for PEX5 recycling (PubMed:[16314507](#), PubMed:[16854980](#), PubMed:[21362118](#), PubMed:[29884772](#)). Specifically recognizes PEX5 monoubiquitinated at 'Cys-11', and pulls it out of the peroxisome lumen through the PEX2-PEX10-PEX12 retrotranslocation channel (PubMed:[29884772](#)). Extraction by the PEX1-PEX6 AAA ATPase complex is accompanied by unfolding of the TPR repeats and release of bound cargo from PEX5 (PubMed:[29884772](#)).

Cellular Location

Cytoplasm, cytosol. Peroxisome membrane. Cell projection, cilium, photoreceptor outer segment. Note=Associated with peroxisomal membranes; anchored by PEX26 to peroxisome membranes (PubMed:12717447, PubMed:16854980). Localized at the base of the outer segment of photoreceptor cells (PubMed:26593283)

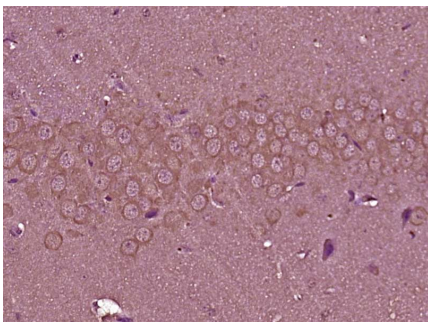
Tissue Location

Expressed in the retina, at higher levels in the photoreceptor layer at the joint between the outer and inner segments

Background

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Images



Paraformaldehyde-fixed, paraffin embedded (Rat brain); 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 (PEX6) Polyclonal Antibody, Unconjugated (AP54907) at 1:400 overnight at 4°C, followed by operating according to SP Kit(Rabbit) (sp-0023) instructions and DAB staining.

Please note: All products are 'FOR RESEARCH USE ONLY. NOT FOR USE IN DIAGNOSTIC OR THERAPEUTIC PROCEDURES'.