

Rabbit Anti-PEX7/AP Conjugated antibody

SL19913R-AP

Product Name	Anti-PEX7/AP
Chinese Name	碱性磷酸酶（AP）标记的过氧化物酶体生成蛋白 7 抗体
Alias	PBD9B; PCDP1; Peroxin 7; Peroxin-7; Peroxisomal PTS2 receptor; Peroxisomal targeting signal 2 receptor; Peroxisome biogenesis factor 7; Peroxisome targeting signal 2 receptor; PEX7; PEX7 protein; PEX7_HUMAN; PTS2 receptor; PTS2R; RCDP1; RD.
Research Area	Cell biology Neurobiology Signal transduction
Immunogen Species	Rabbit
Clonality	Polyclonal
React Species	Mouse(predicted:Human,Rat,Dog,Pig,Cow,Horse,Rabbit,Sheep) WB=1:500-2000
Applications	not yet tested in other applications. optimal dilutions/concentrations should be determined by the end user.
Molecular weight	36kDa
Form	Lyophilized or Liquid
Concentration	1mg/ml
immunogen	KLH conjugated synthetic peptide derived from human PEX7
Lsotype	IgG
Purification	affinity purified by Protein A
Storage Buffer	1M TBS(pH7.4) with 1% BSA, 3% Proclin300 and 50% Glycerol. Store at -20 °C for one year. Avoid repeated freeze/thaw cycles. The lyophilized antibody is stable at room temperature for at least one month and for greater than a year when kept at -20°C. When reconstituted in sterile pH 7.4 1M PBS or diluent of antibody the antibody is stable for at least two weeks at 2-4 °C.
Storage	
Product Detail	background: This gene encodes the cytosolic receptor for the set of peroxisomal matrix enzymes targeted to the organelle by the peroxisome targeting signal 2 (PTS2). Defects in this gene cause peroxisome biogenesis disorders (PBDs), which are characterized by multiple defects in peroxisome function. There are at least 14 complementation groups for PBDs, with more than one 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 have been associated with PBD complementation group 11 (PBD-CG11) disorders, rhizomelic chondrodysplasia punctata type 1 (RCDP1), and Refsum disease (RD). [provided by RefSeq, Oct 2008]

Function:

Binds to the N-terminal PTS2-type peroxisomal targeting signal and plays an essential role in peroxisomal protein import.

Subunit:

nteracts with PEX5.

Subcellular Location:

Peroxisome. Cytoplasm.

Tissue Specificity:

Ubiquitous. Highest expression in pancreas, skeletal muscle and heart.

DISEASE:

Defects in PEX7 are the cause of peroxisome biogenesis disorder complementation group 11 (PBD-CG11) [MIM:614879]. 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 13 distinct genetic groups as concluded from complementation studies. Defects in PEX7 are the cause of rhizomelic chondrodysplasia punctata type 1 (RCDP1) [MIM:215100]. RCDP1 is characterized by rhizomelic shortening of femur and humerus, vertebral disorders, cataract, cutaneous lesions and severe mental retardation.

Defects in PEX7 are the cause of peroxisome biogenesis disorder 9B (PBD9B) [MIM:614879]. A peroxisome biogenesis disorder with unusually mild clinical and biochemical manifestations. Affected individuals manifest a variable phenotype similar to, and in some cases indistinguishable from, classic Refsum disease. Variable features include ocular abnormalities, sensorimotor neuropathy, ichthyosis, deafness, chondrodysplasia punctata without rhizomelia or growth failure.

Similarity:

Belongs to the WD repeat peroxin-7 family.
Contains 6 WD repeats.

Database links:

[Entrez Gene: 5191](#) Human

[Entrez Gene: 18634](#) Mouse

[Entrez Gene: 308718](#) Rat

[Omim: 601757](#) Human

[SwissProt: O00628](#) Human

[SwissProt: P97865](#) Mouse

[Unigene: 280932](#) Human

[Unigene: 338363](#) Mouse

Important Note:

This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.