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[Marker Primary Antibody]

Rabbit Anti-PEX7/Biotin Conjugated antibody

Catalog Number: SL19913R-Bio

FOR RESEARCH USE ONLY

[DATASHEET]

Host:	Rabbit
Isotype:	IgG
Clonality:	Polyclonal
Target:	Anti-PEX7/Biotin
Immunogen:	KLH conjugated synthetic peptide derived from human PEX7
Purification:	affinity purified by Protein A
Concentration:	1mg/ml
Storage:	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.
Applications:	WB=1:500-2000 not yet tested in other applications. optimal dilutions/concentrations should be determined by the end user.
Reactivity:	Mouse(predicted:Human,Rat,Dog,Pig,Cow,Horse,Rabbit,Sheep)
Background:	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.

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

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	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.