Product Name: Peroxin 7 Rabbit Polyclonal Antibody
Catalog #: APRab15989

Summary

Production Name	Peroxin 7 Rabbit Polyclonal Antibody
Description	Rabbit Polyclonal Antibody
Host	Rabbit
Application	IHC,WB,ELISA
Reactivity	Human,Mouse,Rat

Performance

Conjugation	Unconjugated
Modification	Unmodified
Isotype	IgG
Clonality	Polyclonal
Form	Liquid
Storage	Store at 4°C short term. Aliquot and store at -20°C long term. Avoid freeze/thaw cycles.
Buffer	Liquid in PBS containing 50% glycerol, 0.5% BSA and 0.02% New type preservative N.
Purification	Affinity purification

Immunogen

Gene Name	PEX7
Alternative Names	PEX7; PTS2R; Peroxisomal targeting signal 2 receptor; PTS2 receptor; Peroxin-7
Gene ID	5191.0
SwissProt ID	O00628.The antiserum was produced against synthesized peptide derived from human PEX7. AA range:204-253

Application

Dilution Ratio	WB 1:500 - 1:2000. IHC 1:100 - 1:300. ELISA: 1:40000..
Molecular Weight	40kD

Background

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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],disease:Defects in PEX7 are a cause of Refsum disease (RD) [MIM:266500]; also known as phytanic acid oxidase deficiency. RD is clinically characterized by a tetrad of abnormalities: retinitis pigmentosa, peripheral neuropathy, cerebellar ataxia, and elevated protein levels in the cerebrospinal fluid (CSF). Patients exhibit accumulation of the branched-chain fatty acid, phytanic acid, in blood and tissues. Less constant features are nerve deafness, anosmia, skeletal abnormalities, ichthyosis, cataracts and cardiac impairment. Manifestations of the disease appear in the second or third decade of life.,disease:Defects in PEX7 are the cause of peroxisome biogenesis disorder complementation group 11 (PBD-CG11) [MIM:601757]. 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.,disease: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.,function:Binds to the N-terminal PTS2-type peroxisomal targeting signal and plays an essential role in peroxisomal protein import.,similarity:Belongs to the WD repeat peroxin-7 family.,similarity:Contains 6 WD repeats.,subunit:Interacts with PEX5.,tissue specificity:Ubiquitous. Highest expression in pancreas, skeletal muscle and heart.,

Research Area

Image Data

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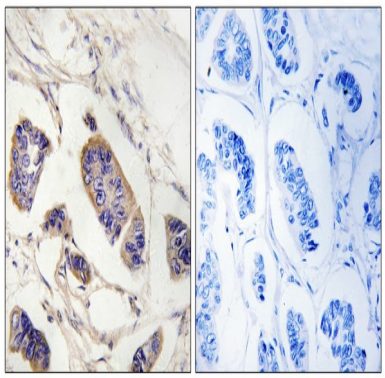
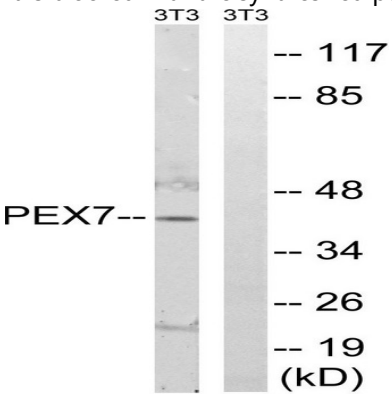
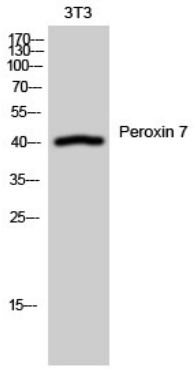


Immunohistochemistry analysis of paraffin-embedded human breast carcinoma tissue, using PEX7 Antibody. The picture on the right is blocked with the synthesized peptide.



Western blot analysis of lysates from NIH/3T3 cells, using PEX7 Antibody. The lane on the right is blocked with the synthesized peptide.



Western Blot analysis of 3T3 cells using Peroxin 7 Polyclonal Antibody

Note

For research use only.