

产品名称: Peroxin 7 Rabbit Polyclonal Antibody
产品货号: AP Rab15989



产品概述 (Summary)

产品名称 (Production Name)	Peroxin 7 Rabbit Polyclonal Antibody
描述 (Description)	Rabbit polyclonal Antibody
宿主 (Host)	Rabbit
应用 (Application)	WB,IHC,ICC/IF,ELISA
种属反应性 (Reactivity)	Human,Mouse,Rat

产品性能 (Performance)

偶联物 (Conjugation)	Unconjugated
修饰 (Modification)	Unmodified
同种型 (Isotype)	IgG
克隆 (Clonality)	Polyclonal
形式 (Form)	Liquid
存放说明 (Storage)	Store at 4°C short term. Aliquot and store at -20°C long term. Avoid freeze/thaw cycles.
储存溶液 (Buffer)	Liquid in PBS containing 50% glycerol, 0.5% protective protein and 0.02% New type preservative N.
纯化方式 (Purification)	Affinity purification

免疫原信息 (Immunogen)

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

产品应用 (Application)

稀释比 (Dilution Ratio)	WB 1:500-1:2000,IHC 1:100-1:300,ICC/IF 1:50-1:200,ELISA 1:20000-1:40000
蛋白分子量 (Molecular Weight)	40kDa

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研究背景 (Background)

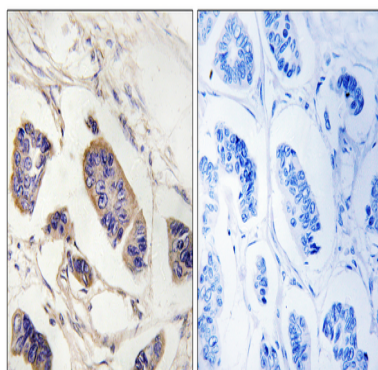
This gene encodes the cytosolic receptor for the set of peroxisomal matrix enzymes targeted to the organelle by the peroxisome targeting signal 2 (PTS2). Defects in this gene cause peroxisome biogenesis disorders (PBDs), which are characterized by multiple defects in peroxisome function. There are at 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], 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.,

研究领域 (Research Area)

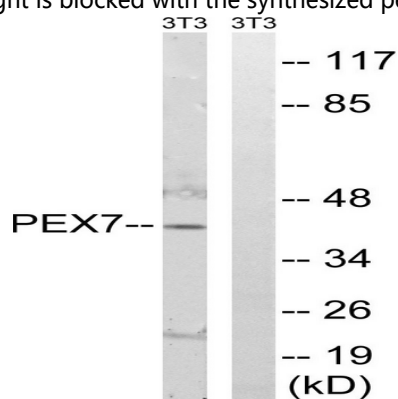
Signal Transduction; Protein Trafficking; Vesicle Transport; Coat Proteins

图片 (Image Data)

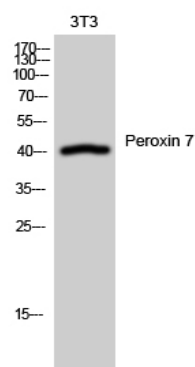
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Immunohistochemistry analysis of paraffin-embedded human breast carcinoma tissue, using PEX7 Antibody. The picture on the right is blocked with the synthesized peptide.



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

注意事项 (Note)

For research use only .