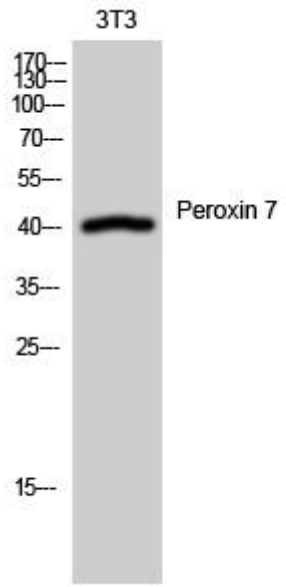
Peroxin 7 Monoclonal Antibody

Catalog No	BYmab-04074
Isotype	IgG
Reactivity	Human;Mouse;Rat
Applications	WB
Gene Name	PEX7
Protein Name	Peroxisomal targeting signal 2 receptor
Immunogen	The antiserum was produced against synthesized peptide derived from human PEX7. AA range:204-253
Specificity	Peroxin 7 Monoclonal Antibody detects endogenous levels of Peroxin 7 protein.
Formulation	Liquid in PBS containing 50% glycerol, 0.5% BSA and 0.02% sodium azide.
Source	Monoclonal, Mouse,IgG
Purification	The antibody was affinity-purified from mouse antiserum by affinity-chromatography using epitope-specific immunogen.
Dilution	WB 1:500-2000
Concentration	1 mg/ml
Purity	≥90%
Storage Stability	-20°C/1 year
Synonyms	PEX7; PTS2R; Peroxisomal targeting signal 2 receptor; PTS2 receptor; Peroxin-7
Observed Band	40kD
Cell Pathway	Peroxisome . Cytoplasm .
Tissue Specificity	Ubiquitous. Highest expression in pancreas, skeletal muscle and heart.
Function	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 disorde
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],
matters needing attention	Avoid repeated freezing and thawing!
Usage suggestions	This product can be used in immunological reaction related experiments. For more information, please consult technical personnel.

Products Images



Western Blot analysis of various cells using Peroxin 7 Monoclonal Antibody