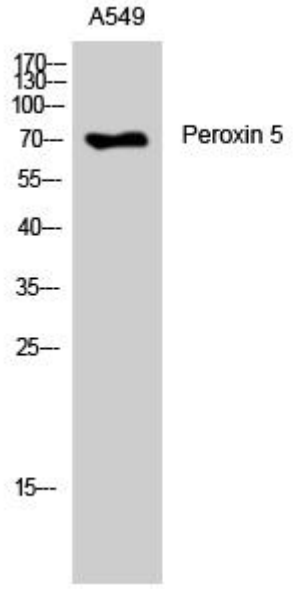
Peroxin 5 Monoclonal Antibody

Catalog No	YP-mAb-04073
Isotype	IgG
Reactivity	Human;Mouse
Applications	WB
Gene Name	PEX5
Protein Name	Peroxisomal targeting signal 1 receptor
Immunogen	Synthesized peptide derived from Peroxin 5 . at AA range: 540-620
Specificity	Peroxin 5 Monoclonal Antibody detects endogenous levels of Peroxin 5 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 1:500-1:2000
Concentration	1 mg/ml
Purity	≥90%
Storage Stability	-20°C/1 year
Synonyms	PEX5; PXR1; Peroxisomal targeting signal 1 receptor; PTS1 receptor; PTS1R; PTS1-BP; Peroxin-5; Peroxisomal C-terminal targeting signal import receptor; Peroxisome receptor 1
Observed Band	70kD
Cell Pathway	Cytoplasm . Peroxisome membrane ; Peripheral membrane protein. Its distribution appears to be dynamic. It is probably a cycling receptor found mainly in the cytoplasm and as well associated to the peroxisomal membrane through a docking factor (PEX13).
Tissue Specificity	Detected in heart, brain, placenta, lung, liver, skeletal muscle, kidney and pancreas.
Function	disease:Defects in PEX5 are a cause of adrenoleukodystrophy neonatal (NALD) [MIM:202370]. NALD is a peroxisome biogenesis disorder (PBD) characterized by the accumulation of very long-chain fatty acids, adrenal insufficiency and mental retardation. Inheritance is autosomal recessive.,disease:Defects in PEX5 are a cause of Zellweger syndrome (ZWS) [MIM:214100]. ZWS is a fatal peroxisome biogenesis disorder characterized by dysmorphic facial features, hepatomegaly, ocular abnormalities, renal cysts, hearing impairment, profound psychomotor retardation, severe hypotonia and neonatal seizures. Death occurs within the first year of life.,disease:Defects in PEX5 may be a cause of infantile Refsum disease (IRD) [MIM:266510]. IRD is a mild peroxisome biogenesis disorder (PBD). Clinical features include early onset, mental retardation, minor facial dysmorphism,

retinopathy, sensorineural hearing

Background	The product of this gene binds to the C-terminal PTS1-type tripeptide peroxisomal targeting signal (SKL-type) and plays an essential role in peroxisomal protein import. Peroxins (PEXs) are proteins that are essential for the assembly of functional peroxisomes. The peroxisome biogenesis disorders (PBDs) are a group of genetically heterogeneous autosomal recessive, lethal diseases characterized by multiple defects in peroxisome function. The peroxisomal biogenesis disorders are a heterogeneous group with at least 14 complementation groups and with more than 1 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 are a cause of neonatal adrenoleukodystrophy (NALD)
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 5 Monoclonal Antibody