



# Peroxin 5 Monoclonal Antibody

<b>Catalog No</b>	BYmab-04073
<b>Isotype</b>	IgG
<b>Reactivity</b>	Human;Mouse
<b>Applications</b>	WB
<b>Gene Name</b>	PEX5
<b>Protein Name</b>	Peroxisomal targeting signal 1 receptor
<b>Immunogen</b>	Synthesized peptide derived from Peroxin 5 . at AA range: 540-620
<b>Specificity</b>	Peroxin 5 Monoclonal Antibody detects endogenous levels of Peroxin 5 protein.
<b>Formulation</b>	Liquid in PBS containing 50% glycerol, 0.5% BSA and 0.02% sodium azide.
<b>Source</b>	Monoclonal, Mouse,IgG
<b>Purification</b>	The antibody was affinity-purified from mouse antiserum by affinity-chromatography using epitope-specific immunogen.
<b>Dilution</b>	WB 1:500-2000
<b>Concentration</b>	1 mg/ml
<b>Purity</b>	≥90%
<b>Storage Stability</b>	-20°C/1 year
<b>Synonyms</b>	PEX5; PXR1; Peroxisomal targeting signal 1 receptor; PTS1 receptor; PTS1R; PTS1-BP; Peroxin-5; Peroxisomal C-terminal targeting signal import receptor; Peroxisome receptor 1
<b>Observed Band</b>	70kD
<b>Cell Pathway</b>	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).
<b>Tissue Specificity</b>	Detected in heart, brain, placenta, lung, liver, skeletal muscle, kidney and pancreas.
<b>Function</b>	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

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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)

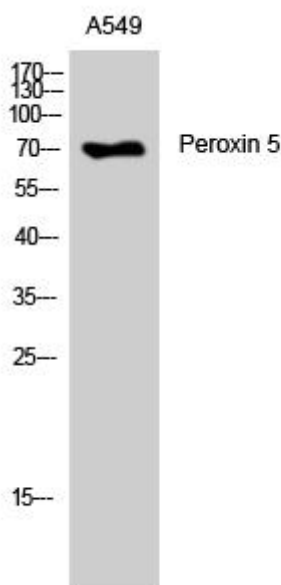
## matters needing attention

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

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