



Peroxin 19 Monoclonal Antibody

Catalog No	YP-mAb-04070
Isotype	IgG
Reactivity	Human;Mouse;Rat
Applications	WB
Gene Name	PEX19
Immunogen	The antiserum was produced against synthesized peptide derived from human PEX19. AA range:219-268
Specificity	Peroxin 19 Monoclonal Antibody detects endogenous levels of Peroxin 19 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	Peroxin 19;PEX19;PMP1;PMPI;PXF;PXMP1;Peroxisomal biogenesis factor 19; HK33;OK/SW-cl.22;33 kDa housekeeping protein;Peroxin-19;Peroxisomal farnesylated protein
Observed Band	33kD
Calculated Molecular Weight	33kD
Cell Pathway	Cytoplasm . Peroxisome membrane ; Lipid-anchor ; Cytoplasmic side . Mainly cytoplasmic. Some fraction membrane-associated to the outer surface of peroxisomes. .
Tissue Specificity	Ubiquitously expressed. Isoform 1 is strongly predominant in all tissues except in utero where isoform 2 is the main form.
Function	alternative products:Experimental confirmation may be lacking for some isoforms,disease:Defects in PEX19 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 PEX19 are the cause of peroxisome biogenesis disorder complementation



group 14 (PBD-CG14) [MIM:600279]; also known as PBD-CGJ. 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 (CRD).

Background

peroxisomal biogenesis factor 19 (PEX19) Homo sapiens This gene is necessary for early peroxisomal biogenesis. It acts both as a cytosolic chaperone and as an import receptor for peroxisomal membrane proteins (PMPs). Peroxisins (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. These disorders have at least 14 complementation groups, with more than one phenotype being observed for some 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 Zellweger syndrome (ZWS), as well as peroxisome biogenesis disorder complementation group 14 (PBD-CG14), which is

matters needing attention

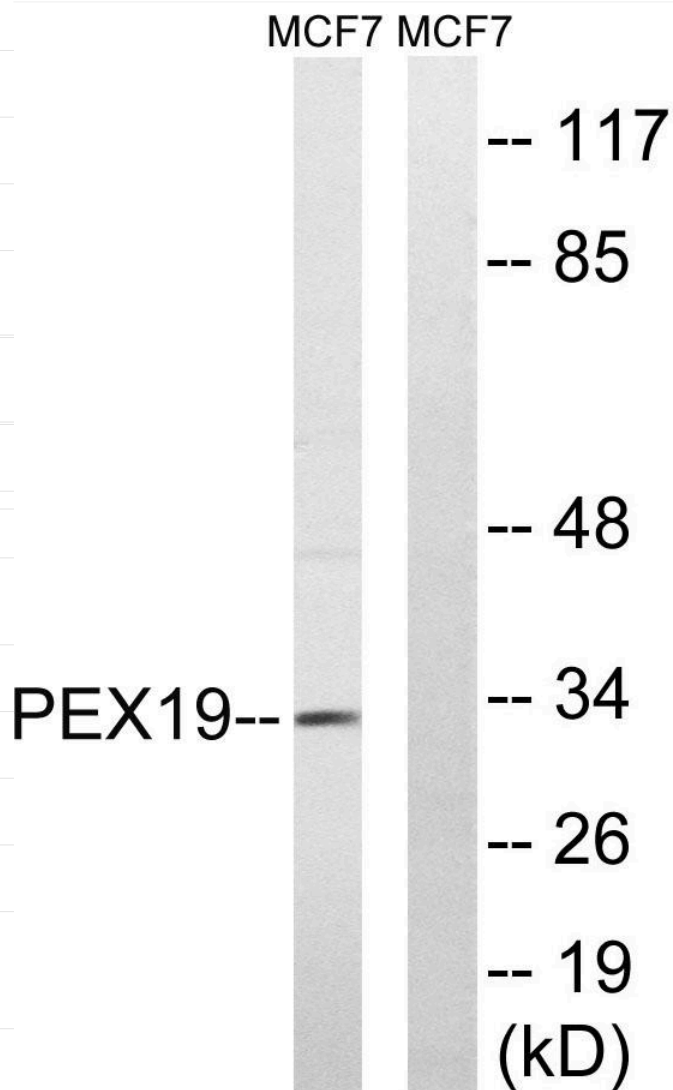
Avoid repeated freezing and thawing!

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 19 Monoclonal Antibody