PEX6 Polyclonal Antibody

Catalog No	YP-Ab-05911
Isotype	IgG
Reactivity	Human;Rat;Mouse
Applications	WB;ELISA
Gene Name	PEX6 PXAAA1
Protein Name	Peroxisome assembly factor 2 (PAF-2) (Peroxin-6) (Peroxisomal biogenesis factor 6) (Peroxisomal-type ATPase 1)
Immunogen	Synthesized peptide derived from human protein . at AA range: 480-560
Specificity	PEX6 Polyclonal Antibody detects endogenous levels of protein.
Formulation	Liquid in PBS containing 50% glycerol, and 0.02% sodium azide.
Source	Polyclonal, Rabbit,IgG
Purification	The antibody was affinity-purified from rabbit antiserum by affinity-chromatography using epitope-specific immunogen.
Dilution	WB 1:500-2000 ELISA 1:5000-20000
Concentration	1 mg/ml
Purity	≥90%
Storage Stability	-20°C/1 year
Synonyms	
Observed Band	107kD
Cell Pathway	Cytoplasm. Peroxisome membrane . Cell projection, cilium, photoreceptor outer segment . Associated with peroxisomal membranes. Localized at the base of the outer segment of photoreceptor cells (PubMed:26593283). .
Tissue Specificity	Expressed in the retina, at higher levels in the photoreceptor layer at the joint between the outer and inner segments.
Function	disease:Defects in PEX6 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 PEX6 are the cause of peroxisome biogenesis disorder complementation group 4 (PBD-CG4) [MIM:601498]; also known as PBD-CGC. 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 cont

Background	This gene encodes a member of the AAA (ATPases associated with diverse cellular activities) family of ATPases. This member is a predominantly cytoplasmic protein, which plays a direct role in peroxisomal protein import and is required for PTS1 (peroxisomal targeting signal 1, a C-terminal tripeptide of the sequence ser-lys-leu) receptor activity. Mutations in this gene cause peroxisome biogenesis disorders of complementation group 4 and complementation group 6. Several transcript variants encoding different isoforms have been found for this gene. [provided by RefSeq, Oct 2015],
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.

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