PEX6 Rabbit pAb

CatalogNo: YN1038

Key Features

Host Species
- Rabbit

Reactivity
- Human,Rat,Mouse

Applications
- WB,ELISA

MW
- 107kD (Observed)

Isotype
- IgG

Recommended Dilution Ratios

WB 1:500-2000
ELISA 1:5000-20000

Storage

Storage*	-15°C to -25°C/1 year(Do not lower than -25°C)
Formulation	Liquid in PBS containing 50% glycerol, and 0.02% sodium azide.

Basic Information

Clonality	Polyclonal

Immunogen Information

Immunogen	Synthesized peptide derived from human protein . at AA range: 480-560
Specificity	PEX6 Polyclonal Antibody detects endogenous levels of protein.

Target Information

Gene name	PEX6 PXAAA1

Protein Name Peroxisome assembly factor 2 (PAF-2) (Peroxin-6) (Peroxisomal biogenesis factor 6) (Peroxisomal-type ATPase 1)

Organism	Gene ID	UniProt ID
Human	5190;	Q13608;
Mouse		Q99LC9;
Rat		P54777;

Cellular Localization 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 continuum of overlapping phenotypes known as the Zellweger spectrum. The PBD group is genetically heterogeneous with at least 14 distinct genetic groups as concluded from complementation studies.,Function:Involved in peroxisome biosynthesis. Required for stability of the PTS1 receptor. Anchored by PEX26 to peroxisome membranes, possibly to form heteromeric AAA ATPase complexes required for the import of proteins into peroxisomes.,similarity:Belongs to the AAA ATPase family.,subcellular location:Associated with peroxisomal membranes.,subunit:Interacts directly with PEX26 and PEX1. Mediates the indirect interaction between PEX1 and PEX26.,

Validation Data

Contact information

Orders: order.cn@immunoway.com
Support: support.cn@immunoway.com
Telephone: 400-8787-807(China)
Website: http://www.immunoway.com.cn
Address: 2200 Ringwood Ave San Jose, CA 95131 USA

Please scan the QR code to access additional product information:
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