

Product name:	Peroxin 3 Rabbit Polyclonal Antibody
Cat number:	ABN15987
Conjugate:	Unconjugated
Size:	100μL
Clone:	Polyclonal
Concentration:	1mg/ml
Host:	Rabbit
Isotype:	IgG
Immunogen:	The antiserum was produced against synthesized peptide derived from human PEX3. AA range:12-61
Reactivity:	Human,Mouse,Rat
Applications:	WB 1:500-1:2000,IHC 1:50-1:300,ELISA 1:2000-1:20000
Molecular Weight:	42kDa
Purification:	Affinity purification
Form:	Liquid
Buffer:	Liquid in PBS containing 50% glycerol, 0.5% BSA and 0.02% New type preservative N.
Storage:	Store at 4°C short term. Aliquot and store at -20°C for 12 months. Avoid freeze/thaw cycles.

Background:

The product of this gene is involved in peroxisome biosynthesis and integrity. It assembles membrane vesicles before the matrix proteins are translocated. 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 Zellweger syndrome (ZWS). [provided by RefSeq, Oct 20] disease:Defects in PEX3 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 PEX3 are the cause of peroxisome biogenesis disorder complementation group 12 (PBD-CG12) [MIM:603164]; also known as PBD-CGG. 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 and integrity. Assembles membrane vesicles before the matrix proteins are translocated. As a docking factor for PEX19, is necessary for the import of peroxisomal membrane proteins in the peroxisomes.,similarity:Belongs to the peroxin-3 family.,subunit:Interacts with PEX19.,tissue specificity:Found in all examined tissues.,