

Product name:	Peroxin 2 Rabbit Polyclonal Antibody
Cat number:	ABN15986
Conjugate:	Unconjugated
Size:	100µL
Clone:	Polyclonal
Concentration:	1mg/ml
Host:	Rabbit
Isotype:	IgG
Immunogen:	The antiserum was produced against synthesized peptide derived from human PXMP3. AA range:1-50
Reactivity:	Human,Rat,Mouse
Applications:	WB 1:500-1:2000,IHC 1:100-1:300,ICC/IF 1:50-1:200,ELISA 1:10000-1:20000
Molecular Weight:	35kDa
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:

This gene encodes an integral peroxisomal membrane protein required for peroxisome biogenesis. The protein is thought to be involved in peroxisomal matrix protein import. Mutations in this gene result in one form of Zellweger syndrome and infantile Refsum disease. Alternative splicing results in multiple transcript variants encoding the same protein. [provided by RefSeq, Jul 2008],disease:Defects in PXMP3 are 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 deficit, hepatomegaly, osteoporosis, failure to thrive, and hypocholesterolemia. The biochemical abnormalities include accumulation of phytanic acid, very long chain fatty acids (VLCFA), di- and trihydroxycholestanic acid and pipercolic acid.,disease:Defects in PXMP3 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 PXMP3 are the cause of peroxisome biogenesis disorder complementation group 5 (PBD-CG5) [MIM:170993]; also known as PBD-CGF. 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:Somewhat implicated in the biogenesis of peroxisomes.,similarity:Belongs to the pex2/pex10/pex12 family.,similarity:Contains 1 RING-type zinc finger.,