

过氧化物酶体生物合成因子 19 抗体

产品货号： mlR12623

英文名称： PEX19

中文名称： 过氧化物酶体生物合成因子 19 抗体

别名： 33 kDa housekeeping protein; D1S2223E; HK33; Housekeeping gene 33kD; OK/SW-cl.22; PBD12A; Peroxin 19; Peroxin-19; Peroxisomal biogenesis factor 19; Peroxisomal farnesylated protein; PEX19; PEX19_HUMAN; PMP1; PMPI; PXF; PXMP1.

研究领域： 细胞生物 信号转导

抗体来源： Rabbit

克隆类型： Polyclonal

交叉反应： Human, Mouse, Rat, Dog, Pig, Cow, Horse,

产品应用： ELISA=1:500-1000 IHC-P=1:400-800 IHC-F=1:400-800 ICC=1:100-500 IF=1:100-500 （石蜡切片需做抗原修复）

not yet tested in other applications.

optimal dilutions/concentrations should be determined by the end user.

分子量： 33kDa

细胞定位： 细胞浆

性状： Lyophilized or Liquid

浓度： 1mg/ml

免疫原 : KLH conjugated synthetic peptide derived from human PEX19:201-299/299

亚型 : IgG

纯化方法 : affinity purified by Protein A

储存液 : 0.01M TBS(pH7.4) with 1% BSA, 0.03% Proclin300 and 50% Glycerol.

保存条件 : Store at -20 ° C for one year. Avoid repeated freeze/thaw cycles. The lyophilized antibody is stable at room temperature for at least one month and for greater than a year when kept at -20° C. When reconstituted in sterile pH 7.4 0.01M PBS or diluent of antibody the antibody is stable for at least two weeks at 2-4 ° C.

PubMed : PubMed

产品介绍 : 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 also known as PBD-CGJ. Alternative splicing results in multiple transcript variants. [provided by RefSeq, Aug 2010]

Function:

Necessary for early peroxisomal biogenesis. Acts both as a cytosolic chaperone and as an import receptor for peroxisomal membrane proteins (PMPs). Binds and stabilizes newly synthesized PMPs in the cytoplasm by interacting with their hydrophobic membrane-spanning domains, and targets them to the peroxisome membrane by binding to the integral membrane protein PEX3. Excludes CDKN2A from the nucleus and prevents its interaction with MDM2, which results in active degradation of TP53.

Subcellular Location:

Cytoplasm. Peroxisome membrane. 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.

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 (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.

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.

Similarity:

Belongs to the peroxin-19 family.

SWISS:

P40855

Gene ID:

5824

Important Note:

This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.