

本公司提供的电子版本说明书仅供参考，实验请以收到的纸质手册为准。

Anti-Human/Mouse PEX5 Polyclonal Antibody

多克隆抗体

产品货号: PA05555

第三版

描述:The product of this gene binds to the C-terminal PTS1-type tripeptide peroxisomal targeting signal (SKL-type) and plays an essential role in peroxisomal protein import. 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. 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 of neonatal adrenoleukodystrophy (NALD), a cause of Zellweger syndrome (ZWS) as well as may be a cause of infantile Refsum disease (IRD). Alternatively spliced transcript variants encoding different isoforms have been identified.

抗原:Synthesized peptide derived from the C-terminal region of human Peroxin 5

配方:

如何使用:加1ml超纯水重溶

稳定性: -20 ° C保存条件下，冻干粉,保质期为五年；液体，保质期为两年。

稀释液:PBS (pH7.4) ， 1% BSA

应用:WB 1 ~ 5 μ g/ml.

特异性:Detected in heart, brain, placenta, lung, liver, skeletal muscle, kidney and pancreas.