abcam

Product datasheet

Anti-MPV17 antibody ab67466

1 图像

概述

产品名称	Anti-MPV17抗体
描述	**小鼠多克隆抗体**to MPV17
宿主	Mouse
经测试应用	**适用于:** WB
种属反应性	**与反应:** Human
免疫原	Full length protein (Human)

性能

形式	Liquid
存放说明	Shipped at 4°C. Upon delivery aliquot and store at -20°C or -80°C. Avoid repeated freeze / thaw cycles.
存储溶液	Preservative: None Constituents: PBS, pH 7.2
纯度	Protein A purified
克隆	**多克隆**
同种型	IgG

应用

Our Abpromise guarantee covers the use of **ab67466** in the following tested applications.

The application notes include recommended starting dilutions; optimal dilutions/concentrations should be determined by the end user.

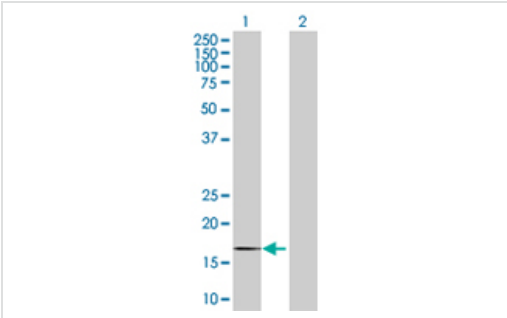
应用	Ab评论	说明
WB		1/500 - 1/1000. Detects a band of approximately 17 kDa (predicted molecular weight: 20 kDa).

靶标

功能	Involved in mitochondria homeostasis. May be involved in the metabolism of reactive oxygen

	species and control of oxidative phosphorylation and mitochondrial DNA (mtDNA) maintenance.
组织特异性	Ubiquitous. Expressed in pancreas, kidney, muscle, liver, lung, placenta, brain and heart.
疾病相关	Defects in MPV17 are a cause of hepatocerebral mitochondrial DNA depletion syndrome (MDS) [MIM:251880]. MDS is a clinically heterogeneous group of disorders characterized by a reduction in mitochondrial DNA (mtDNA) copy number. Primary mtDNA depletion is inherited as an autosomal recessive trait and may affect single organs, typically muscle or liver, or multiple tissues. Individuals with the hepatocerebral form of mitochondrial DNA depletion syndrome have early progressive liver failure and neurologic abnormalities, hypoglycemia, and increased lactate in body fluids. Defects in MPV17 are the cause of Navajo neurohepatopathy (NN) [MIM:256810]. NN is an autosomal recessive disease that is prevalent among Navajo children in the southwestern United States. The major clinical features are hepatopathy, peripheral neuropathy, corneal anesthesia and scarring, acral mutilation, cerebral leukoencephalopathy, failure to thrive, and recurrent metabolic acidosis with intercurrent infections. Infantile, childhood, and classic forms of NN have been described. Mitochondrial DNA depletion was detected in the livers of patients, suggesting a primary defect in mtDNA maintenance.
序列相似性	Belongs to the peroxisomal membrane protein PXMP2/4 family.
细胞定位	Mitochondrion inner membrane.

图片



Western blot - MPV17 antibody (ab67466)

All lanes : Anti-MPV17 antibody (ab67466) at 1/500 dilution

Lane 1 : MPV17 transfected 293T cell lysate
Lane 2 : Non-transfected 293T cell lysate

Lysates/proteins at 25 µg/ml per lane.

Secondary
All lanes : Goat Anti-Mouse IgG (H&L)-HRP Conjugate at 1/2500 dilution

Predicted band size: 20 kDa
Observed band size: 17 kDa

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