abcam

Product datasheet

Anti-ATP7A antibody ab42486

2 References 2 图像

概述

产品名称	Anti-ATP7A抗体
描述	**兔多克隆抗体**to ATP7A
宿主	Rabbit
经测试应用	**适用于:** ICC/IF, WB, IHC-P
种属反应性	**与反应:** Human **预测可用于:** Mouse, Cat, Dog, Chimpanzee
免疫原	Synthetic peptide corresponding to Human ATP7A. Short peptide sequence used to raise this antibody is 100% homologous to isoform 4 (1500aa, 163kDa), 1 (1514aa, 165kDa), 2 (1581aa, 172kDa), 5 (1422aa, 154kDa) of human ATP7A Database link: Q04656
阳性对照	WB: HepG2 cell lysate. IHC-P: Human kidney

性能

形式	Liquid
存放说明	Shipped at 4°C. Upon delivery aliquot and store at -20°C. Avoid freeze / thaw cycles.
存储溶液	Constituents: PBS, 2% Sucrose
纯度	Immunogen affinity purified
克隆	**多克隆**
同种型	IgG

应用

Our Abpromise guarantee covers the use of **ab42486** 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
ICC/IF		Use a concentration of 5 µg/ml.

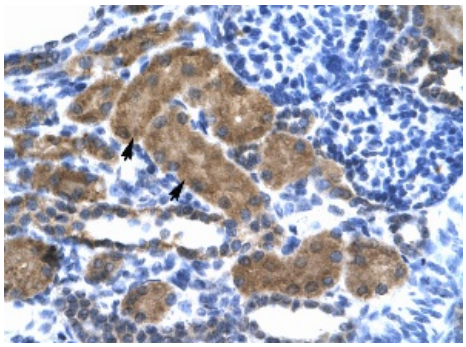
应用	Ab评论	说明
WB		Use a concentration of 0.5 µg/ml. Predicted molecular weight: 30 kDa. Good results were obtained when blocked with 5% non-fat dry milk in 0.05% PBS-T.
IHC-P		Use at an assay dependent concentration.

靶标

功能	May supply copper to copper-requiring proteins within the secretory pathway, when localized in the trans-Golgi network. Under conditions of elevated extracellular copper, it relocalized to the plasma membrane where it functions in the efflux of copper from cells.
组织特异性	Found in most tissues except liver. Isoform 3 is widely expressed including in liver cell lines. Isoform 1 is expressed in fibroblasts, choriocarcinoma, colon carcinoma and neuroblastoma cell lines. Isoform 2 is expressed in fibroblasts, colon carcinoma and neuroblastoma cell lines.
疾病相关	Defects in ATP7A are the cause of Menkes disease (MNKD) [MIM:309400]; also known as kinky hair disease. MNKD is an X-linked recessive disorder of copper metabolism characterized by generalized copper deficiency. MNKD results in progressive neurodegeneration and connective-tissue disturbances: focal cerebral and cerebellar degeneration, early growth retardation, peculiar hair, hypopigmentation, cutis laxa, vascular complications and death in early childhood. The clinical features result from the dysfunction of several copper-dependent enzymes. Defects in ATP7A are the cause of occipital horn syndrome (OHS) [MIM:304150]; also known as X-linked cutis laxa. OHS is an X-linked recessive disorder of copper metabolism. Common features are unusual facial appearance, skeletal abnormalities, chronic diarrhea and genitourinary defects. The skeletal abnormalities included occipital horns, short, broad clavicles, deformed radii, ulnae and humeri, narrowing of the rib cage, undercalcified long bones with thin cortical walls and coxa valga. Defects in ATP7A are a cause of distal spinal muscular atrophy X-linked type 3 (DSMAX3) [MIM:300489]. DSMAX3 is a neuromuscular disorder. Distal spinal muscular atrophy, also known as distal hereditary motor neuronopathy, represents a heterogeneous group of neuromuscular disorders caused by selective degeneration of motor neurons in the anterior horn of the spinal cord, without sensory deficit in the posterior horn. The overall clinical picture consists of a classical distal muscular atrophy syndrome in the legs without clinical sensory loss. The disease starts with weakness and wasting of distal muscles of the anterior tibial and peroneal compartments of the legs. Later on, weakness and atrophy may expand to the proximal muscles of the lower limbs and/or to the distal upper limbs.
序列相似性	Belongs to the cation transport ATPase (P-type) (TC 3.A.3) family. Type IB subfamily. Contains 6 HMA domains.
结构域	The C-terminal di-leucine, 1487-Leu-Leu-1488, is an endocytic targeting signal which functions in retrieving recycling from the plasma membrane to the TGN. Mutation of the di-leucine signal results in the accumulation of the protein in the plasma membrane.
细胞定位	Endoplasmic reticulum; Cytoplasm > cytosol and Golgi apparatus > trans-Golgi network membrane. Cell membrane. Cycles constitutively between the trans-Golgi network (TGN) and the plasma membrane. Predominantly found in the TGN and relocalized to the plasma membrane in response to elevated copper levels.

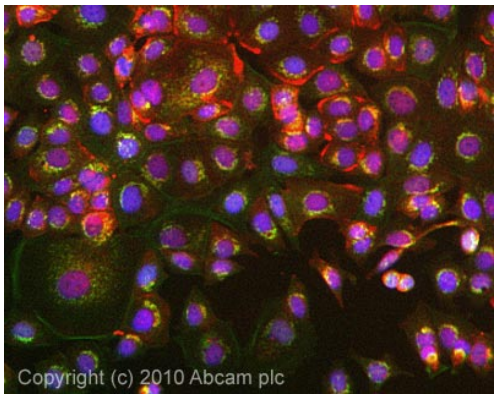
图片

图片



Immunohistochemistry (Formalin/PFA-fixed paraffin-embedded sections) - Anti-ATP7A antibody (ab42486)

Antibody ab42486 (4.0-8.0 ug/ml) immunohistochemistry staining of paraffin embedded human kidney. Positive staining of epithelial cells of human renal tubules idicated by arrows.



Immunocytochemistry/ Immunofluorescence - Anti-ATP7A antibody (ab42486)

ICC/IF image of ab42486 stained Mcf7 cells. The cells were 4% formaldehyde fixed (10 min) and then incubated in 1%BSA / 10% normal goat serum / 0.3M glycine in 0.1% PBS-Tween for 1h to permeabilise the cells and block non-specific protein-protein interactions. The cells were then incubated with the antibody (ab42486, 5µg/ml) overnight at +4°C. The secondary antibody (green) was Alexa Fluor® 488 goat anti-rabbit IgG (H+L) used at a 1/1000 dilution for 1h. Alexa Fluor® 594 WGA was used to label plasma membranes (red) at a 1/200 dilution for 1h. DAPI was used to stain the cell nuclei (blue) at a concentration of 1.43µM.

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