产品名称: ATP7A Rabbit Polyclonal Antibody
产品货号: APRab07343

产品概述（Summary）

描述(Description)	Rabbit polyclonal Antibody
宿主(Host)	Rabbit
应用(Application)	IHC,ICC/IF,ELISA
种属反应(Reactivity)	Human,Mouse,Rat
偶联物(Conjugation)	Unconjugated
修饰(Modification)	Unmodified
同种型(Isotype)	IgG
克隆(Clonality)	Polyclonal
剂型(Form)	Liquid
保存条件(Storage)	Store at 4°C short term. Aliquot and store at -20°C long term. Avoid freeze/thaw cycles.
储存溶液(Buffer)	Liquid in PBS containing 50% glycerol, 0.5% protective protein and 0.02% New type preservative N.
纯化 (Purification)	Affinity purification

产品应用(Application)

稀释比(Dilution Ratio)	IHC 1:100-1:300,ICC/IF 1:50-1:200,ELISA 1:20000-1:40000
分子量(Molecular Weight)	

抗原信息(Antigen Information)

基因名(Gene Name)	ATP7A
别名(Alternative Names)	ATP7A; MC1; MNK; Copper-transporting ATPase 1; Copper pump 1; Menkes disease-associated protein
基因 ID(Gene ID)	538.0
SwissProt ID	Q04656
免疫原(Immunogen)	The antiserum was produced against synthesized peptide derived from human ATP7A. AA range:591-640

研究背景（Background）

ATPase copper transporting alpha(ATP7A) Homo sapiens This gene encodes a transmembrane protein that functions in

copper transport across membranes. This protein is localized to the trans Golgi network, where it is predicted to supply copper to copper-dependent enzymes in the secretory pathway. It relocalizes to the plasma membrane under conditions of elevated extracellular copper, and functions in the efflux of copper from cells. Mutations in this gene are associated with Menkes disease, X-linked distal spinal muscular atrophy, and occipital horn syndrome. Alternatively-spliced transcript variants have been observed. [provided by RefSeq, Aug 2013],catalytic activity:ATP + H(2)O + Cu(2+)(In) = ADP + phosphate + Cu(2+)(Out).,disease: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.,disease: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.,domain: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.,function: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.,online information:Heavy metal - Issue 79 of February 2007,similarity:Belongs to the cation transport ATPase (P-type) family. Type IB subfamily.,similarity:Contains 6 HMA domains.,subcellular location: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.,subunit:Monomer.,tissue specificity: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.,

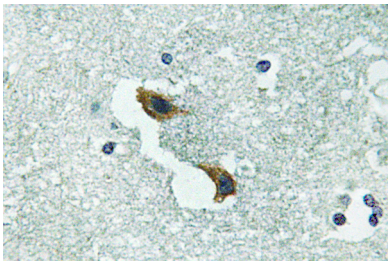
研究领域（Research Area）

Angiogenesis; MAPK

注意事项（Note）

For research use only.

图片（Image Data）



Immunohistochemistry analysis of ATP7A antibody in paraffin-embedded human brain tissue.

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产品类别	产品货号	产品名称
WB 解决方案	RA10020	2h 极速 WB 即用型全流程试剂盒
	RA10021	4h 快速 WB 即用型全流程试剂盒
	RA10042	5h 畅享版 WB 全流程试剂盒
	RA10037	校准级彩色预染蛋白 Marker（8-180kDa）
	RA10038	校准级彩色预染蛋白 Marker（10-250kDa）
	RA10039	校准级高分子彩色预染蛋白 Marker（25-400kDa）
TSA 多重荧光染色试剂盒	RA10008	TSA 双标三色多重荧光染色试剂盒（mIHC）
	RA10009	TSA 三标四色多重荧光染色试剂盒（mIHC）
	RA10010	TSA 四标五色多重荧光染色试剂盒（mIHC）
	RA10011	TSA 五标六色多重荧光染色试剂盒（mIHC）
	RA10012	TSA 六标七色多重荧光染色试剂盒（mIHC）
IHC 检测试剂盒	RA10006	HRP Anti-Mouse/Rabbit IHC Detection System
	RA10007	Polymer-HRP Anti-Mouse/Rabbit IHC Detection System
抗体标记试剂盒	RE80004p	辣根过氧化物酶(HRP)抗体标记试剂盒
	RE80002q	Sulfo-NHS-生物素标记试剂盒
	RE80007p	Cy3 荧光素标记试剂盒
	RE80011p	Fluor488 荧光素标记试剂盒
	RE80017p	Fluor750 荧光素标记试剂盒
	RE80005p	藻红蛋白(R-PE) 抗体快速标记试剂盒
	RE80040	PE-Cy7 串联染料抗体快速标记试剂盒
稳转细胞系构建服务 (免费赠送全膜 WB 验证)	TS-0001	过表达稳转细胞系构建
	TS-0002	敲低稳转细胞系构建
	TS-0003	敲除细胞系构建

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