Product Name: ATP7A Rabbit Polyclonal Antibody
Catalog #: APRab07343

Summary

Production Name	ATP7A Rabbit Polyclonal Antibody
Description	Rabbit Polyclonal Antibody
Host	Rabbit
Application	IHC-P,IF-P,IF-F,ICC/IF,ELISA
Reactivity	Human,Mouse,Rat

Performance

Conjugation	Unconjugated
Modification	Unmodified
Isotype	IgG
Clonality	Polyclonal
Form	Liquid
Storage	Store at 4°C short term. Aliquot and store at -20°C long term. Avoid freeze/thaw cycles.
Buffer	Liquid in PBS containing 50% glycerol, 0.5% protective protein and 0.02% New type preservative N.
Purification	Affinity purification

Immunogen

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

Application

Dilution Ratio	IHC-P 1:100-1:300, ELISA 1:40000, IF-P/IF-F/ICC/IF 1:50-200
Molecular Weight	

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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.,

Research Area

Angiogenesis; MAPK

Image Data

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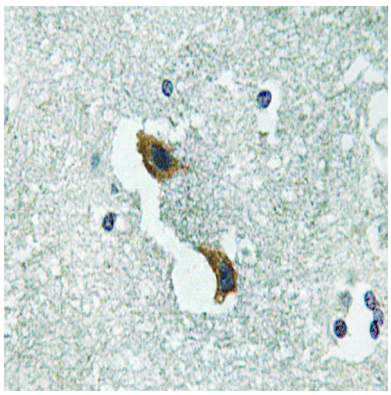

Immunohistochemistry analysis of ATP7A antibody in paraffin-embedded human brain tissue.

Note

For research use only.