

Rabbit Anti-ATP7A antibody

SL1572R

Product Name:	ATP7A
Chinese Name:	铜Transporter质α链抗体
Alias:	ATP 7A; ATPase Copper Transporting Alpha Polypeptide; ATPase Cu++ transporting alpha polypeptide (Menkes syndrome); ATPase Cu++ transporting alpha polypeptide; Copper pump 1; Copper transporting ATPase 1; Cu++ transporting P type ATPase; MC 1; MC1; Menkes disease-associated protein; Menkes syndrome; MK; MNK; OHS; ATP7A_HUMAN.
	Specific References(1) SL1572R has been referenced in 1 publications.
文献引用	[IF=2.09] Wang, Xurui, et al. "miR-133a enhances the sensitivity of Hep-2 cells and
Pub	vincristine-resistant Hep-2v cells to cisplatin by downregulating ATP7B expression."
	International Journal of Molecular Medicine. 37(6):1636-42.other;Others.
	PubMed:27121102
Organism Species:	Rabbit
Clonality:	Polyclonal
React Species:	Human,Mouse,Rat,Dog,Cow,Horse,Rabbit,
Applications:	ELISA=1:500-1000IHC-P=1:400-800IHC-F=1:400-800IF=1:100-500 (Paraffin sections need antigen repair) not yet tested in other applications. optimal dilutions/concentrations should be determined by the end user.
Molecular weight:	163kDa
Cellular localization:	cytoplasmicThe cell membrane
Form:	Lyophilized or Liquid
Concentration:	1mg/ml
immunogen:	KLH conjugated synthetic peptide derived from human ATP7A:242- 285/1500 <cytoplasmic></cytoplasmic>
Lsotype:	IgG
Purification:	affinity purified by Protein A

Storage Buffer:	0.01M TBS(pH7.4) with 1% BSA, 0.03% Proclin300 and 50% Glycerol.
	Store at -20 °C for one year. Avoid repeated freeze/thaw cycles. The lyophilized
Storage:	antibody is stable at room temperature for at least one month and for greater than a year
	when kept at -20°C. When reconstituted in sterile pH 7.4 0.01M PBS or diluent of
	antibody the antibody is stable for at least two weeks at 2-4 °C.
PubMed:	PubMed
Product Detail:	Copper-transporting ATPase 1 is an integral membrane protein cycling constitutively between the trans-golgi network and the plasma membrane. It 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. Defects in ATP7A are the cause of Menkes syndrome; also known as kinky hair disease, an X-linked recessive disorder. 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. Subunit: Monomer. Interacts with PDZD11. Subcellular Location: Golgi apparatus. trans-Golgi network membrane; Multi-pass membrane protein. Cell membrane; Multi-pass membrane protein. Note: 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. Isoform 3: Cytoplasm. cytosol. Isoform 5: Endoplasmic reticulum. Tissue Specificity: Found in most fissues 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. DISEASE: Menkes discase (MNKD) [MIM:309400]: 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. A mild form of the disease has been described, in which cerebellar ataxia and

Occipital horn syndrome (OHS) [MIM:304150]: An X-linked recessive disorder of copper metabolism. Common features are unusual facial appearance, skeletal abnormalities, chronic diarrhea and genitourinary defects. The skeletal abnormalities include 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. Note=The disease is caused by mutations affecting the gene represented in this entry.

Distal spinal muscular atrophy, X-linked, 3 (DSMAX3) [MIM:300489]: 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. Note=The disease is caused by mutations affecting the gene represented in this entry.

Similarity:

Belongs to the cation transport ATPase (P-type) (TC 3.A.3) family. Type IB subfamily. Contains 6 HMA domains.

SWISS: Q04656

Gene ID: 538

Database links:

Entrez Gene: 538Human

Entrez Gene: 11977Mouse

Entrez Gene: 24941Rat

<u>Omim: 300011</u>Human

SwissProt: Q04656Human

SwissProt: Q64430Mouse

SwissProt: P70705Rat

Unigene: 496414Human

Unigene: 254297Mouse

Unigene: 10554Rat





