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Rabbit Anti-ATP7A/Cy5 Conjugated antibody

SL1572R-Cy5

Product Name	Anti-ATP7A/Cy5
Chinese Name	Cy5 标记的铜 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.
Research Area	immunology Channel protein Transporter
Immunogen Species	Rabbit
Clonality	Polyclonal
React Species	Human,Mouse,Rat(predicted:Dog,Cow,Horse,Rabbit)
Applications	Flow-Cyt=2ug/Test,IF=1:100-500 not yet tested in other applications. optimal dilutions/concentrations should be determined by the end user.
Molecular weight	163kDa
Form	Lyophilized or Liquid
Concentration	1mg/ml
immunogen	KLH conjugated synthetic peptide derived from human ATP7A C-terminus
Lsotype	IgG
Purification	affinity purified by Protein A
Storage Buffer	1M TBS(pH7.4) with 1% BSA, 3% Proclin300 and 50% Glycerol.
Storage	Store at -20 °C for one year. Avoid repeated freeze/thaw cycles. The lyophilized 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 1M PBS or diluent of antibody the antibody is stable for at least two weeks at 2-4 °C.
Product Detail	**background:** 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 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.

DISEASE:
Menkes disease (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 moderate developmental delay predominate. Note=The disease is caused by mutations affecting the gene represented in this entry.
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.

Database links:

Entrez Gene: 538 Human

Entrez Gene: 11977 Mouse

Entrez Gene: 24941 Rat

Omim: 300011 Human

SwissProt: Q04656 Human

SwissProt: Q64430 Mouse

SwissProt: P70705 Rat

Unigene: 496414 Human

Unigene: 254297 Mouse

Unigene: 10554 Rat

Important Note:
This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.