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Product datasheet

Anti-ATP7A antibody [L60/4] ab131400

3 References 画像数 2

製品の概要

製品名	Anti-ATP7A antibody [L60/4]
製品の詳細	Mouse monoclonal [L60/4] to ATP7A
由来種	Mouse
アプリケーション	**適用あり:** ICC/IF, Flow Cyt
種交差性	**交差種:** Mouse, Human **交差が予測される動物種:** Chinese hamster
免疫原	Synthetic peptide corresponding to Human ATP7A aa 42-61. Sequence: SLEEKNATIIYDPKLQTPKT Database link: **Q04656** Run BLAST with Run BLAST with
ポジティブ・コントロール	ICC/IF: NIH/3T3 cells. Flow Cyt: HT1080 cells.
特記事項	The clone number has been updated from S60-4 to L60/4, both clone numbers name the same antibody clone. The Life Science industry has been in the grips of a reproducibility crisis for a number of years. Abcam is leading the way in addressing this with our range of recombinant monoclonal antibodies and knockout edited cell lines for gold-standard validation. Please check that this product meets your needs before purchasing. If you have any questions, special requirements or concerns, please send us an inquiry and/or contact our Support team ahead of purchase. Recommended alternatives for this product can be found below, along with publications, customer reviews and Q&As

製品の特性

製品の状態	Liquid
保存方法	Shipped at 4°C. Store at -20°C.
バッファー	Preservative: 0.09% Sodium azide Constituents: PBS, 50% Glycerol (glycerin, glycerine)
精製度	Protein G purified

ポリ/モノ	モノクローナル
クローン名	L60/4
アイソタイプ	IgG2b

アプリケーション

The Abpromise guarantee **Abpromise保証は、** 次のテスト済みアプリケーションにおけるab131400の使用に適用されます

アプリケーションノートには、推奨の開始希釈率がありますが、適切な希釈率につきましてはご検討ください。

アプリケーション	Abreviews	特記事項
ICC/IF		1/100.
Flow Cyt		Use 0.1µg for 10^6 cells. **ab170192** - Mouse monoclonal IgG2b, is suitable for use as an isotype control with this antibody.

ターゲット情報

機能

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.

組織特異性

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.

関連疾患

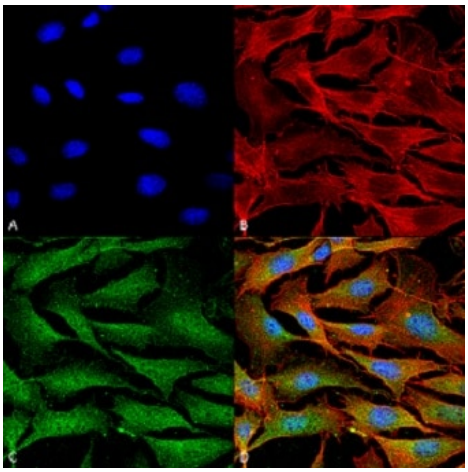
Defects in ATP7A are the cause of Menkes disease (MNKD) [MIM:309400]; also known as kinky hair disease. MNKD is an X-linked recessive disorder of copper metabolism characterized by generalized copper deficiency. MNKD results in progressive neurodegeneration and connective-tissue disturbances: focal cerebral 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.

配列類似性

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

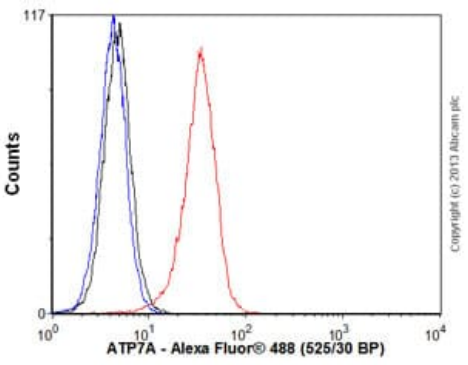
	Contains 6 HMA domains.
ドメイン	The C-terminal di-leucine, 1487-Leu-Leu-1488, is an endocytic targeting signal which functions in retrieving recycling from the plasma membrane to the TGN. Mutation of the di-leucine signal results in the accumulation of the protein in the plasma membrane.
細胞内局在	Endoplasmic 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.

画像



Immunocytochemistry/ Immunofluorescence - Anti-ATP7A antibody [L60/4] (ab131400)

NIH/3T3 (Mouse embryo fibroblast cell line) cells labeling ATP7A using ab131400 at 1/100 dilution in ICC/IF. Cells were fixed using 4% formaldehyde for 15 minutes at room temperature. Incubation with primary antibody was performed for 1 hour at room temperature. Secondary antibody used was a goat anti-mouse ATTO 488 (green) at 1/200 dilution for 1 hour at room temperature. Counterstained with Phalloidin Texas Red F-actin stain. Nuclei were stained with DAPI (blue).



Flow Cytometry - Anti-ATP7A antibody [L60/4] (ab131400)

Overlay histogram showing HT1080 cells stained with ab131400 (red line). The cells were fixed with 4% paraformaldehyde (10 min) and then permeabilized with 0.1% PBS-Tween for 20 min. The cells were then incubated in 1x PBS / 10% normal goat serum / 0.3M glycine to block non-specific protein-protein interactions followed by the antibody (ab131400, 0.1µg/1x10^6 cells) for 30 min at 22°C. The secondary antibody used was Alexa Fluor® 488 goat anti-mouse IgG (H+L) (**ab150113**) at 1/2000 dilution for 30 min at 22°C. Isotype control antibody (black line) was mouse IgG2b [PLPV219] (**ab91366**, 1µg/1x10^6 cells) used under the same conditions. Unlabelled sample (blue line) was also used as a control. Acquisition of >5,000 events were collected using a 20mW Argon ion laser (488nm) and 525/30 bandpass filter. This antibody gave a positive signal in HT1080 cells fixed with 80% methanol (5 min)/permeabilized with 0.1% PBS-Tween for 20 min used under the same conditions.

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