abcam

Product datasheet

Anti-CTNS antibody ab220391

1 Image

Overview

Product name	Anti-CTNS antibody
Description	Rabbit polyclonal to CTNS
Host species	Rabbit
Tested applications	**Suitable for:** ICC/IF
Species reactivity	**Reacts with:** Human **Predicted to work with:** Mouse, Rat, Cow
Immunogen	Recombinant fragment corresponding to Human CTNS aa 250-350. Database link: **O60931** Run BLAST with Run BLAST with
Positive control	A431 cells.
General notes	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

Properties

Form	Liquid
Storage instructions	Shipped at 4°C. Store at +4°C short term (1-2 weeks). Upon delivery aliquot. Store at -20°C long term. Avoid freeze / thaw cycle.
Storage buffer	pH: 7.20 Preservative: 0.02% Sodium azide Constituents: 40% Glycerol (glycerin, glycerine), 59% PBS
Purity	Immunogen affinity purified
Clonality	Polyclonal
Isotype	IgG

Applications

The Abpromise guarantee Our **Abpromise guarantee** covers the use of ab220391 in the following tested applications.

The application notes include recommended starting dilutions; optimal dilutions/concentrations should be determined by the end user.

Application	Abreviews	Notes
ICC/IF		Use a concentration of 0.25 - 2 µg/ml.

Target

Function Thought to transport cystine out of lysosomes.

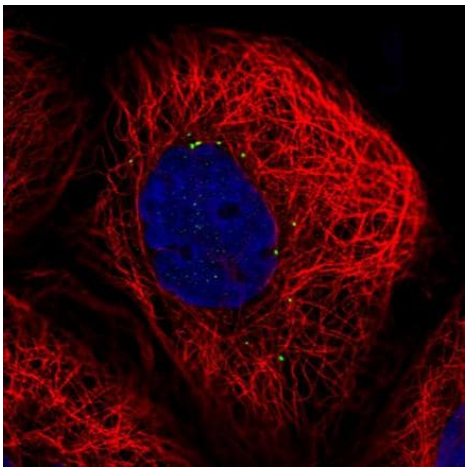
Tissue specificity Strongly expressed in pancreas, kidney (adult and fetal) and in skeletal muscle. Expressed at lower levels in placenta and heart. Weakly expressed in lung, liver and brain (adult and fetal).

Involvement in disease Cystinosis, nephropathic type (CTNS) [MIM:219800]: A form of cystinosis, a lysosomal storage disease due to defective transport of cystine across the lysosomal membrane. This results in cystine accumulation and crystallization in the cells causing widespread tissue damage. The classical nephropathic form has onset in the first year of life and is characterized by a polyuro-polydipsic syndrome, marked height-weight growth delay, generalized impaired proximal tubular reabsorptive capacity, with severe fluid-electrolyte balance alterations, renal failure, ocular symptoms and other systemic complications. Note=The disease is caused by mutations affecting the gene represented in this entry.
Cystinosis, adult, non-nephropathic type (CTNSANN) [MIM:219750]: A form of cystinosis, a lysosomal storage disease due to defective transport of cystine across the lysosomal membrane. This results in cystine accumulation and crystallization in the cells causing widespread tissue damage. Cystinosis adult non-nephropathic type is characterized by ocular features and a benigne course. Patients manifest mild photophobia due to conjunctival and corneal cystine crystals. Note=The disease is caused by mutations affecting the gene represented in this entry.
Cystinosis, late-onset juvenile or adolescent nephropathic type (CTNSJAN) [MIM:219900]: A form of cystinosis, a lysosomal storage disease due to defective transport of cystine across the lysosomal membrane. This results in cystine accumulation and crystallization in the cells causing widespread tissue damage. Late-onset juvenile or adolescent nephropathic cystinosis is an intermediated form, manifesting first at age 10 to 12 years with proteinuria due to glomerular damage rather than with the manifestations of tubular damage that occur first in infantile cystinosis. There is no excess amino aciduria and stature is normal. Photophobia, late development of pigmentary retinopathy, and chronic headaches are features. Note=The disease is caused by mutations affecting the gene represented in this entry.

Sequence similarities Belongs to the cystinosin family.
Contains 2 PQ-loop domains.

Cellular localization Lysosome membrane.

Images



Immunocytochemistry/ Immunofluorescence - Anti-CTNS antibody (ab220391)

Immunofluorescent analysis of PFA-fixed, Triton X-100 permeabilized A431 cells labeling CTNS with ab220391 at 4 µg/ml (green).

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- We provide support in Chinese, English, French, German, Japanese and Spanish
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