SANTA CRUZ BIOTECHNOLOGY, INC.

citrin (H-69): sc-98624

BACKGROUND

Citrin, also known as SLC25A13 (solute carrier family 25 member 13), ARALAR2 or CTLN2, is a 675 amino acid multi-pass membrane protein that localizes to the inner membrane of the mitochondrion. Expressed in liver, pancreas, kidney, brain, heart and placenta, citrin functions as a calcium-dependent glutamate and aspartate carrier that is thought to play a role in the urea cycle. Citrin, a member of the mitochondrial carrier family, contains three solcar repeats and four EF-hand domains through which it binds calcium. Defects in the gene encoding citrin are the cause of citrullinemia type 2 (CTLN2) and neonatal intrahepatic cholestasis due to citrin deficiency (NICCD). CTLN2 is an autosomal recessive disease that results from errors in the urea cycle and is characterized by neuropsychiatric symptoms such as loss of memory, seizures and coma. NICCD, a non-lethal disorder, occurs during infancy and is characterized by low birth weight, reduced bile flow, growth retardation and hepatic fibrosis.

REFERENCES

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2. Kobayashi, K., et al. 1999. The gene mutated in adult-onset type II citrullinaemia encodes a putative mitochondrial carrier protein. Nat. Genet. 22: 159-163.
3. Del Arco, A., et al. 2000. Characterization of a second member of the subfamily of calcium-binding mitochondrial carriers expressed in human non-excitable tissues. Biochem. J. 345: 725-732.
4. Palmieri, L., et al. 2001. Citrin and ARALAR1 are Ca^{2+}-stimulated aspartate/glutamate transporters in mitochondria. EMBO J. 20: 5060-5069.
5. Yamaguchi, N., et al. 2002. Screening of SLC25A13 mutations in early and late onset patients with citrin deficiency and in the Japanese population: Identification of two novel mutations and establishment of multiple DNA diagnosis methods for nine mutations. Hum. Mutat. 19: 122-130.
6. Saheki, T. and Kobayashi, K. 2002. Mitochondrial aspartate glutamate carrier (citrin) deficiency as the cause of adult-onset type II citrullinemia (CTLN2) and idiopathic neonatal hepatitis (NICCD). J. Hum. Genet. 47: 333-341.
7. Lu, Y.B., et al. 2005. Frequency and distribution in East Asia of 12 mutations identified in the SLC25A13 gene of Japanese patients with citrin deficiency. J. Hum. Genet. 50: 338-346.
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CHROMOSOMAL LOCATION

Genetic locus: SLC25A13 (human) mapping to 7q21.3; Slc25a13 (mouse) mapping to 6 A1.

SOURCE

citrin (H-69) is a rabbit polyclonal antibody raised against amino acids 1-68 mapping at the N-terminus of citrin of human origin.

PRODUCT

Each vial contains 200 µg IgG in 1.0 ml of PBS with < 0.1% sodium azide and 0.1% gelatin.

APPLICATIONS

citrin (H-69) is recommended for detection of citrin of mouse and human origin by Western Blotting (starting dilution 1:200, dilution range 1:100-1:1000), immunoprecipitation [1-2 µg per 100-500 µg of total protein (1 ml of cell lysate)], immunofluorescence (starting dilution 1:50, dilution range 1:50-1:500) and solid phase ELISA (starting dilution 1:30, dilution range 1:30-1:3000).

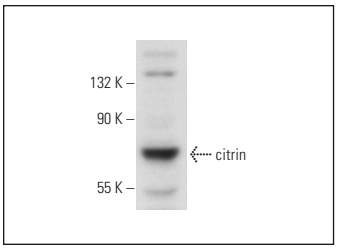
citrin (H-69) is also recommended for detection of citrin in additional species, including canine and porcine.

Suitable for use as control antibody for citrin siRNA (h): sc-89601, citrin siRNA (m): sc-105206, citrin shRNA Plasmid (h): sc-89601-SH, citrin shRNA Plasmid (m): sc-105206-SH, citrin shRNA (h) Lentiviral Particles: sc-89601-V and citrin shRNA (m) Lentiviral Particles: sc-105206-V.

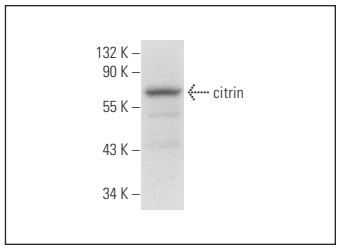
Molecular Weight of citrin: 74 kDa.

Positive Controls: Hep G2 cell lysate: sc-2227 or human liver extract: sc-363766.

DATA



citrin (H-69): sc-98624. Western blot analysis of citrin expression in Hep G2 whole cell lysate.



citrin (H-69): sc-98624. Western blot analysis of citrin expression in human liver tissue extract.

STORAGE

Store at 4° C, **DO NOT FREEZE**. Stable for one year from the date of shipment. Non-hazardous. No MSDS required.

RESEARCH USE

For research use only, not for use in diagnostic procedures.