

## citrin (G-12): sc-104144

### 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

1. Sinasac, D.S., Crackower, M.A., Lee, J.R., Kobayashi, K., Saheki, T., Scherer, S.W. and Tsui, L.C. 1999. Genomic structure of the adult-onset type II citrullinemia gene, SLC25A13, and cloning and expression of its mouse homologue. *Genomics* 62: 289-292.
2. Kobayashi, K., Sinasac, D.S., Iijima, M., Boright, A.P., Begum, L., Lee, J.R., Yasuda, T., Ikeda, S., Hirano, R., Terazono, H., Crackower, M.A., Kondo, I., Tsui, L.C., Scherer, S.W. and Saheki, T. 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., Agudo, M. and Satrústegui, J. 2000. Characterization of a second member of the subfamily of calcium-binding mitochondrial carriers expressed in human non-excitabile tissues. *Biochem. J.* 345: 725-732.
4. Palmieri, L., Pardo, B., Lasorsa, F.M., del Arco, A., Kobayashi, K., Iijima, M., Runswick, M.J., Walker, J.E., Saheki, T., Satrústegui, J. and Palmieri, F. 2001. Citrin and ARALAR1 are Ca<sup>2+</sup>-stimulated aspartate/glutamate transporters in mitochondria. *EMBO J.* 20: 5060-5069.
5. Yamaguchi, N., Kobayashi, K., Yasuda, T., Nishi, I., Iijima, M., Nakagawa, M., Osame, M., Kondo, I. and Saheki, T. 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.

### CHROMOSOMAL LOCATION

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

### SOURCE

citrin (G-12) is an affinity purified goat polyclonal antibody raised against a peptide mapping near the N-terminus of citrin of human origin.

### RESEARCH USE

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

### PRODUCT

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

Blocking peptide available for competition studies, sc-104144 P, (100 µg peptide in 0.5 ml PBS containing < 0.1% sodium azide and 0.2% BSA).

### APPLICATIONS

citrin (G-12) is recommended for detection of citrin of mouse, rat 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 (G-12) is also recommended for detection of citrin in additional species, including equine, canine, bovine 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.

### RECOMMENDED SECONDARY REAGENTS

To ensure optimal results, the following support (secondary) reagents are recommended: 1) Western Blotting: use donkey anti-goat IgG-HRP: sc-2020 (dilution range: 1:2000-1:100,000) or Cruz Marker™ compatible donkey anti-goat IgG-HRP: sc-2033 (dilution range: 1:2000-1:5000), Cruz Marker™ Molecular Weight Standards: sc-2035, TBS Blotto A Blocking Reagent: sc-2333 and Western Blotting Luminol Reagent: sc-2048. 2) Immunoprecipitation: use Protein A/G PLUS-Agarose: sc-2003 (0.5 ml agarose/2.0 ml). 3) Immunofluorescence: use donkey anti-goat IgG-FITC: sc-2024 (dilution range: 1:100-1:400) or donkey anti-goat IgG-TR: sc-2783 (dilution range: 1:100-1:400) with UltraCruz™ Mounting Medium: sc-24941.

### STORAGE

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

### PROTOCOLS

See our web site at [www.scbt.com](http://www.scbt.com) or our catalog for detailed protocols and support products.



Try **citrin (D-7): sc-393303** or **citrin (8Z): sc-100937**, our highly recommended monoclonal alternatives to citrin (G-12).