

OTC (T-13): sc-132403

BACKGROUND

OTC (ornithine carbamoyltransferase), also known as OTC_{Case}, is a 354 amino acid protein that belongs to the ATCase/OTCase family of proteins. Expressed in liver and intestinal mucosa, OTC localizes to the mitochondrial matrix and exists as a homotrimer. Specifically, OTC plays a vital role in the urea cycle, catalyzing the second step in this pathway: the formation of L-citrulline from L-ornithine and carbamoyl phosphate. In humans, the urea cycle is an important pathway to detoxification of ammonia. Mutations in the gene encoding OTC are associated with the X-linked disorder OTCD (ornithine carbamoyltransferase deficiency). OTCD is a disorder of the urea cycle characterized by hyperammonemia. In males, OTCD is fatal, whereas females express variable symptoms. In addition, the OTC gene localizes near a region of the X chromosome that is associated with Duchenne muscular dystrophy, suggesting a possible role in that disease as well.

REFERENCES

- Lindgren, V., de Martinville, B., Horwich, A.L., Rosenberg, L.E. and Francke, U. 1984. Human ornithine transcarbamylase locus mapped to band Xp21.1 near the Duchenne muscular dystrophy locus. *Science* 226: 698-700.
- Hata, A., Tsuzuki, T., Shimada, K., Takiguchi, M., Mori, M. and Matsuda, I. 1986. Isolation and characterization of the human ornithine transcarbamylase gene: structure of the 5'-end region. *J. Biochem.* 100: 717-725.
- Tuchman, M., Holzknacht, R.A., Gueron, A.B., Berry, S.A. and Tsai, M.Y. 1992. Six new mutations in the ornithine transcarbamylase gene detected by single-strand conformational polymorphism. *Pediatr. Res.* 32: 600-604.
- Trivedi, M., Zafar, S., Spalding, M.J. and Jonnalagadda, S. 2001. Ornithine transcarbamylase deficiency unmasked because of gastrointestinal bleeding. *J. Clin. Gastroenterol.* 32: 340-343.
- Online Mendelian Inheritance in Man, OMIM™. 2002. Johns Hopkins University, Baltimore, MD. MIM Number: 300461. World Wide Web URL: <http://www.ncbi.nlm.nih.gov/omim/>
- Yamaguchi, S., Brailey, L.L., Morizono, H., Bale, A.E. and Tuchman, M. 2006. Mutations and polymorphisms in the human ornithine transcarbamylase (OTC) gene. *Hum. Mutat.* 27: 626-632.
- Suriano, G., Azevedo, L., Novais, M., Boscolo, B., Seruca, R., Amorim, A. and Ghibaudi, E.M. 2007. *In vitro* demonstration of intra-locus compensation using the ornithine transcarbamylase protein as model. *Hum. Mol. Genet.* 16: 2209-2214.
- Chan, J.S., Harding, C.O. and Blanke, C.D. 2008. Postchemotherapy hyperammonemic encephalopathy emulating ornithine transcarbamoylase (OTC) deficiency. *South. Med. J.* 101: 543-545.

CHROMOSOMAL LOCATION

Genetic locus: OTC (human) mapping to Xp11.4; Otc (mouse) mapping to X A1.1.

SOURCE

OTC (T-13) is an affinity purified goat polyclonal antibody raised against a peptide mapping within an internal region of OTC 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.

Blocking peptide available for competition studies, sc-132403 P, (100 µg peptide in 0.5 ml PBS containing < 0.1% sodium azide and 0.2% BSA).

APPLICATIONS

OTC (T-13) is recommended for detection of OTC of mouse, rat and human origin by Western Blotting (starting dilution 1:200, dilution range 1:100-1:1000), 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).

OTC (T-13) is also recommended for detection of OTC in additional species, including equine, canine, bovine and porcine.

Suitable for use as control antibody for OTC siRNA (h): sc-91306, OTC siRNA (m): sc-151338, OTC shRNA Plasmid (h): sc-91306-SH, OTC shRNA Plasmid (m): sc-151338-SH, OTC shRNA (h) Lentiviral Particles: sc-91306-V and OTC shRNA (m) Lentiviral Particles: sc-151338-V.

Molecular Weight of OTC precursor: 40 kDa.

Molecular Weight of mature OTC subunit of homotrimer: 36 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) 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.

RESEARCH USE

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

PROTOCOLS

See our web site at www.scbt.com or our catalog for detailed protocols and support products.