

# ORNT1/2 (T-20): sc-85088

## BACKGROUND

ORNT1 (mitochondrial ornithine transporter 1), also known as solute carrier family 25 member 15, is a 301 amino acid protein that is localized to the mitochondrial inner membrane. ORNT1 functions to transport ornithine across the inner membrane from the cytoplasm to the mitochondrial matrix, which is an essential step in the urea cycle, the process of eliminating toxic ammonium ions from the breakdown of amino acids. While it is expressed at very low levels in all other tissues, ORNT1 is relatively highly expressed in pancreas and liver. ORNT2 is also known as SLC25A2 and, like ORNT1, is a 301 amino acid protein that plays an essential role in ornithine transport. Defects in the gene encoding ORNT1 are the cause of hyperornithinemia-hyperammonemia-homocitrullinuria syndrome (HHH syndrome), an autosomal recessive disorder characterized by cerebellar ataxia, mental retardation and disturbance in consciousness. These symptoms are a result of defective ornithine transport, which prevents ammonia from being converted to urea and excreted, therefore leading to the buildup of ammonia within the body.

## REFERENCES

1. Camacho, J.A., et al. 1999. Hyperornithinaemia-hyperammonaemia-homocitrullinuria syndrome is caused by mutations in a gene encoding a mitochondrial ornithine transporter. *Nat. Genet.* 22: 151-158.
2. Tsujino, S., et al. 2000. Three novel mutations (G27E, insAAC, R179X) in the ORNT1 gene of Japanese patients with hyperornithinemia, hyperammonemia, and homocitrullinuria syndrome. *Ann. Neurol.* 47: 625-631.
3. Salvi, S., et al. 2001. Seven novel mutations in the ORNT1 gene (SLC25A15) in patients with hyperornithinemia, hyperammonemia, and homocitrullinuria syndrome. *Hum. Mutat.* 18: 460.
4. Salvi, S., et al. 2001. Clinical and molecular findings in hyperornithinemia-hyperammonemia-homocitrullinuria syndrome. *Neurology* 57: 911-914.
5. Miyamoto, T., et al. 2002. A novel mutation, P126R, in a Japanese patient with HHH syndrome. *Pediatr. Neurol.* 26: 65-67.
6. Wan, D., et al. 2004. Large-scale cDNA transfection screening for genes related to cancer development and progression. *Proc. Natl. Acad. Sci. USA* 101: 15724-15729.
7. Online Mendelian Inheritance in Man, OMIM™. 2009. Johns Hopkins University, Baltimore, MD. MIM Number: 238970. World Wide Web URL: <http://www.ncbi.nlm.nih.gov/omim/>

## SOURCE

ORNT1/2 (T-20) is an affinity purified goat polyclonal antibody raised against a peptide mapping within an internal region of ORNT1 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-85088 P, (100 µg peptide in 0.5 ml PBS containing < 0.1% sodium azide and 0.2% BSA).

## APPLICATIONS

ORNT1/2 (T-20) is recommended for detection of ORNT1 and ORNT2 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).

ORNT1/2 (T-20) is also recommended for detection of ORNT1 and ORNT2 in additional species, including bovine and porcine.

Molecular Weight of ORNT1: 33 kDa.

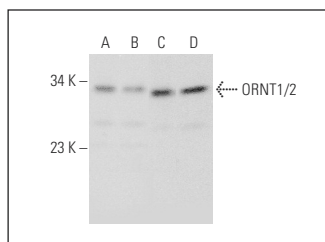
Molecular Weight of ORNT2: 33 kDa.

Positive Controls: MCF7 whole cell lysate: sc-2206, Raji whole cell lysate: sc-364236 or K-562 whole cell lysate: sc-2203.

## 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.

## DATA



ORNT1/2 (T-20): sc-85088. Western blot analysis of ORNT1/2 expression in MCF7 (A), Raji (B), K-562 (C) and Jurkat (D) whole cell lysates.

## 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](http://www.scbt.com) or our catalog for detailed protocols and support products.