



SZABO SCANDIC

Part of Europa Biosite

Produktinformation



Forschungsprodukte & Biochemikalien



Zellkultur & Verbrauchsmaterial



Diagnostik & molekulare Diagnostik



Laborgeräte & Service

Weitere Information auf den folgenden Seiten!
See the following pages for more information!



Lieferung & Zahlungsart

siehe unsere [Liefer- und Versandbedingungen](#)

Zuschläge

- Mindermengenzuschlag
- Trockeneiszuschlag
- Gefahrgutzuschlag
- Expressversand

SZABO-SCANDIC HandelsgmbH

Quellenstraße 110, A-1100 Wien

T. +43(0)1 489 3961-0

F. +43(0)1 489 3961-7

mail@szabo-scandic.com

www.szabo-scandic.com

[linkedin.com/company/szaboscandic](https://www.linkedin.com/company/szaboscandic) 

ORNT1 siRNA (m): sc-151317

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. 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 buildup within the body.

REFERENCES

1. Camacho, J.A., Obie, C., Biery, B., Goodman, B.K., Hu, C.A., Almashanu, S., Steel, G., Casey, R., Lambert, M., Mitchell, G.A. and Valle, D. 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., Kanazawa, N., Ohashi, T., Eto, Y., Saito, T., Kira, J. and Yamada, T. 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., Dionisi-Vici, C., Bertini, E., Verardo, M. and Santorelli, F.M. 2001. Seven novel mutations in the ORNT1 gene (SLC25A15) in patients with hyperornithinemia, hyperammonemia, and homocitrullinuria syndrome. *Hum. Mutat.* 18: 460.
4. Salvi, S., Santorelli, F.M., Bertini, E., Boldrini, R., Meli, C., Donati, A., Burlina, A.B., Rizzo, C., Di Capua, M., Fariello, G. and Dionisi-Vici, C. 2001. Clinical and molecular findings in hyperornithinemia-hyperammonemia-homocitrullinuria syndrome. *Neurology* 57: 911-914.
5. Miyamoto, T., Kanazawa, N., Hayakawa, C. and Tsujino, S. 2002. A novel mutation, P126R, in a Japanese patient with HHH syndrome. *Pediatr. Neurol.* 26: 65-67.
6. Wan, D., Gong, Y., Qin, W., Zhang, P., Li, J., Wei, L., Zhou, X., Li, H., Qiu, X., Zhong, F., He, L., Yu, J., Yao, G., Jiang, H., Qian, L., Yu, Y., Shu, H., Chen, X., Xu, H., Guo, M., Pan, Z., Chen, Y., Ge, C., Yang, S. and Gu, J. 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/>

PROTOCOLS

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

CHROMOSOMAL LOCATION

Genetic locus: Slc25a15 (mouse) mapping to 8 A2.

PRODUCT

ORNT1 siRNA (m) is a pool of 3 target-specific 19-25 nt siRNAs designed to knock down gene expression. Each vial contains 3.3 nmol of lyophilized siRNA, sufficient for a 10 μ M solution once resuspended using protocol below. Suitable for 50-100 transfections. Also see ORNT1 shRNA Plasmid (m): sc-151317-SH and ORNT1 shRNA (m) Lentiviral Particles: sc-151317-V as alternate gene silencing products.

For independent verification of ORNT1 (m) gene silencing results, we also provide the individual siRNA duplex components. Each is available as 3.3 nmol of lyophilized siRNA. These include: sc-151317A, sc-151317B and sc-151317C.

STORAGE AND RESUSPENSION

Store lyophilized siRNA duplex at -20° C with desiccant. Stable for at least one year from the date of shipment. Once resuspended, store at -20° C, avoid contact with RNAses and repeated freeze thaw cycles.

Resuspend lyophilized siRNA duplex in 330 μ l of the RNase-free water provided. Resuspension of the siRNA duplex in 330 μ l of RNase-free water makes a 10 μ M solution in a 10 μ M Tris-HCl, pH 8.0, 20 mM NaCl, 1 mM EDTA buffered solution.

APPLICATIONS

ORNT1 siRNA (m) is recommended for the inhibition of ORNT1 expression in mouse cells.

SUPPORT REAGENTS

For optimal siRNA transfection efficiency, Santa Cruz Biotechnology's siRNA Transfection Reagent: sc-29528 (0.3 ml), siRNA Transfection Medium: sc-36868 (20 ml) and siRNA Dilution Buffer: sc-29527 (1.5 ml) are recommended. Control siRNAs or Fluorescein Conjugated Control siRNAs are available as 10 μ M in 66 μ l. Each contain a scrambled sequence that will not lead to the specific degradation of any known cellular mRNA. Fluorescein Conjugated Control siRNAs include: sc-36869, sc-44239, sc-44240 and sc-44241. Control siRNAs include: sc-37007, sc-44230, sc-44231, sc-44232, sc-44233, sc-44234, sc-44235, sc-44236, sc-44237 and sc-44238.

RT-PCR REAGENTS

Semi-quantitative RT-PCR may be performed to monitor ORNT1 gene expression knockdown using RT-PCR Primer: ORNT1 (m)-PR: sc-151317-PR (20 μ l). Annealing temperature for the primers should be 55-60° C and the extension temperature should be 68-72° C.

RESEARCH USE

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