



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) 

GCH-I (h2): 293T Lysate: sc-159499

BACKGROUND

GTP cyclohydrolase I (GCH-I), a homododecamer, catalyzes the conversion of GTP into dihydroneopterin triphosphate and folate. GCH-I is the first and rate limiting enzyme in tetrahydrobiopterin (BH4) biosynthesis. BH4 is the cofactor for tyrosine hydroxylase, a rate-limiting enzyme for dopamine synthesis and tryptophan hydroxylase, the rate-limiting enzyme for serotonin biosynthesis. Dopamine and Serotonin are neurotransmitters involved in depression, which may be associated with a deficiency of BH4. Mutations in the gene encoding GCH-I cause malignant hyperphenylalaninemia, a genetic neurological disorder characterized by abnormally high levels of serum phenylalanine, and dopa-responsive dystonia (DRD), a group of movement disorders characterized by a progressive difficulty in walking which respond to L-dopa.

REFERENCES

1. Maita, N., Okada, K., Hatakeyama, K. and Hakoshima, T. 2002. Crystal structure of the stimulatory complex of GTP cyclohydrolase I and its feedback regulatory protein GFRP. *Proc. Natl. Acad. Sci. USA* 99: 1212-1217.
2. Basset, G., Quinlivan, E.P., Ziemak, M.J., Diaz De La Garza, R., Fischer, M., Schiffmann, S., Bacher, A., Gregory, J.F. and Hanson, A.D. 2002. Folate synthesis in plants: the first step of the pterin branch is mediated by a unique bimodular GTP cyclohydrolase I. *Proc. Nat. Acad. Sci. USA* 99: 12489-12494.
3. Ishii, M., Shimizu, S., Wajima, T., Hagiwara, T., Negoro, T., Miyazaki, A., Tobe, T. and Kiuchi, Y. 2005. Reduction of GTP cyclohydrolase I feedback regulating protein expression by hydrogen peroxide in vascular endothelial cells. *J. Pharmacol. Sci.* 97: 299-302.
4. Hagenah, J., Saunders-Pullman, R., Hedrich, K., Kabakci, K., Habermann, K., Wieggers, K., Mohrmann, K., Lohnau, T., Raymond, D., Vieregge, P., Nygaard, T., Ozelius, L.J., Bressman, S.B. and Klein, C. 2005. High mutation rate in dopa-responsive dystonia: detection with comprehensive GCHI screening. *Neurology* 64: 908-911.
5. Hui xie, H., Wu, Z.Y., Wang, N. and Murong, S.X. 2005. Gene symbol: GTP cyclohydrolase I (GCH-I). Disease: dopa-responsive dystonia. *Hum. Genet.* 116: 235.
6. Nandi, M., Miller, A., Stidwill, R., Jacques, T.S., Lam, A.A., Haworth, S., Heales, S. and Vallance, P. 2005. Pulmonary hypertension in a GTP-cyclohydrolase 1-deficient mouse. *Circulation* 111: 2086-2090.
7. Kealey, C., Roche, S., Claffey, E. and McKeon, P. 2005. Linkage and candidate gene analysis of 14q22-24 in bipolar disorder: support for GCHI as a novel susceptibility gene. *Am. J. Med. Genet. B Neuropsychiatr. Genet.* 136: 75-80.
8. Christensen, R., Alhonen, L., Wahlfors, J., Jakobsen, M. and Jensen, T.G. 2005. Characterization of transgenic mice with the expression of phenylalanine hydroxylase and GTP cyclohydrolase I in the skin. *Exp. Dermatol.* 14: 535-542.
9. Pieper, G.M., Nilakantan, V., Halligan, N.L., Khanna, A.K., Hilton, G. and Vásquez-Vivar, J. 2005. Nitric oxide formation in acutely rejecting cardiac allografts correlates with GTP cyclohydrolase I activity. *Biochem. J.* 391: 541-547.

CHROMOSOMAL LOCATION

Genetic locus: GCH1 (human) mapping to 14q22.2.

PRODUCT

GCH-I (h2): 293T Lysate represents a lysate of human GCH-I transfected 293T cells and is provided as 100 µg protein in 200 µl SDS-PAGE buffer.

APPLICATIONS

GCH-I (h2): 293T Lysate is suitable as a Western Blotting positive control for human reactive GCH-I antibodies. Recommended use: 10-20 µl per lane.

Control 293T Lysate: sc-117752 is available as a Western Blotting negative control lysate derived from non-transfected 293T cells.

STORAGE

Store at -20° C. Repeated freezing and thawing should be minimized. Sample vial should be boiled once prior to use. 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 for detailed protocols and support products.