

GCH-I (G-8): sc-376483

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.

CHROMOSOMAL LOCATION

Genetic locus: GCH1 (human) mapping to 14q22.2; Gch1 (mouse) mapping to 14 C1.

SOURCE

GCH-I (G-8) is a mouse monoclonal antibody raised against amino acids 1-250 representing full length GCH-I of human origin.

PRODUCT

Each vial contains 200 µg IgG_{2a} kappa light chain in 1.0 ml of PBS with < 0.1% sodium azide and 0.1% gelatin.

GCH-I (G-8) is available conjugated to agarose (sc-376483 AC), 500 µg/0.25 ml agarose in 1 ml, for IP; to HRP (sc-376483 HRP), 200 µg/ml, for WB, IHC(P) and ELISA; to either phycoerythrin (sc-376483 PE), fluorescein (sc-376483 FITC), Alexa Fluor® 488 (sc-376483 AF488), Alexa Fluor® 546 (sc-376483 AF546), Alexa Fluor® 594 (sc-376483 AF594) or Alexa Fluor® 647 (sc-376483 AF647), 200 µg/ml, for WB (RGB), IF, IHC(P) and FCM; and to either Alexa Fluor® 680 (sc-376483 AF680) or Alexa Fluor® 790 (sc-376483 AF790), 200 µg/ml, for Near-Infrared (NIR) WB, IF and FCM.

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APPLICATIONS

GCH-I (G-8) is recommended for detection of GCH-I isoforms GCH-1, GCH-2, GCH-3 and GCH-4 of mouse, rat and human origin by Western Blotting (starting dilution 1:100, 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).

Suitable for use as control antibody for GCH-I siRNA (h): sc-60675, GCH-I siRNA (m): sc-60676, GCH-I shRNA Plasmid (h): sc-60675-SH, GCH-I shRNA Plasmid (m): sc-60676-SH, GCH-I shRNA (h) Lentiviral Particles: sc-60675-V and GCH-I shRNA (m) Lentiviral Particles: sc-60676-V.

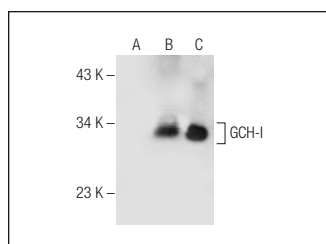
Molecular Weight of GCH-I: 26 kDa.

Positive Controls: IMR-32 cell lysate: sc-2409, SK-N-SH cell lysate: sc-2410 or GCH-I (h): 293T Lysate: sc-159287.

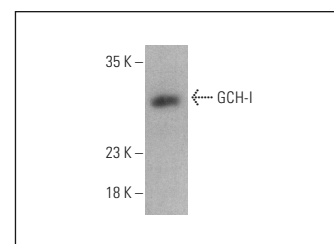
RECOMMENDED SUPPORT REAGENTS

To ensure optimal results, the following support reagents are recommended: 1) Western Blotting: use m-IgGκ BP-HRP: sc-516102 or m-IgGκ BP-HRP (Cruz Marker): sc-516102-CM (dilution range: 1:1000-1:10000), Cruz Marker™ Molecular Weight Standards: sc-2035, UltraCruz® Blocking Reagent: sc-516214 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 m-IgGκ BP-FITC: sc-516140 or m-IgGκ BP-PE: sc-516141 (dilution range: 1:50-1:200) with UltraCruz® Mounting Medium: sc-24941 or UltraCruz® Hard-set Mounting Medium: sc-359850.

DATA



GCH-I (G-8): sc-376483. Western blot analysis of GCH-I expression in non-transfected 293T: sc-117752 (A), human GCH-I transfected 293T: sc-159287 (B) and IMR-32 (C) whole cell lysates.



GCH-I (G-8) HRP: sc-376483 HRP. Direct western blot analysis of GCH-I expression in SK-N-SH whole cell lysate.

SELECT PRODUCT CITATIONS

- Jones, L., et al. 2017. Translational effects and coding potential of an upstream open reading frame associated with DOPA Responsive Dystonia. *Biochim. Biophys. Acta Mol. Basis Dis.* 1863: 1171-1182.
- Nasser, A., et al. 2018. Heterozygous mutations in GTP-cyclohydrolase-1 reduce BH4 biosynthesis but not pain sensitivity. *Pain* 159: 1012-1024.
- Shin, N., et al. 2019. Uncoupled endothelial nitric oxide synthase enhances p-Tau in chronic traumatic encephalopathy mouse model. *Antioxid. Redox Signal.* 30: 1601-1620.
- Sampath, C., et al. 2019. Impairment of Nrf2- and nitroergic-mediated gastrointestinal motility in an MPTP mouse model of Parkinson's disease. *Dig. Dis. Sci.* 64: 3502-3517.
- Lindsay, A., et al. 2021. Tetrahydrobiopterin synthesis and metabolism is impaired in dystrophin-deficient mdx mice and humans. *Acta Physiol.* 231: e13627.

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.