

Product Name: KD-Validated GTP Cyclohydrolase 1 Mouse Monoclonal Antibody**Catalog #: KVA00533**

For research use only.

Summary

Description	KO&KD-Validated antibody
Host	Mouse
Application	WB
Reactivity	Human
Conjugation	Unconjugated
Modification	Unmodified
Isotype	Mouse IgG2a
Clonality	Mouse mAb
Form	Liquid
Storage	Aliquot and store at -20°C (valid for 12 months). Avoid freeze/thaw cycles.
Shipping	Ice bags
Buffer	Supplied in PBS (pH 7.4) containing 50% glycerol, and 0.02% sodium azide.
Purification	Affinity purification

Application

Dilution Ratio	WB 1:500-1:2,500
Molecular Weight	Calculated MW: 27.9kDa

Antigen Information

Gene Name	GCH1 GCH1; GTP Cyclohydrolase 1; GTP Cyclohydrolase I; GTPCH1; DYT5a; DYT5; GCH; Dystonia
Alternative Names	14; EC 3.5.4.16; GTP-CH-I; DYT14; Guanosine 5'-Triphosphate Cyclohydrolase I; Dopa-Responsive Dystonia; GTP-CH-1; HPABH4B
Gene ID	2643.0
SwissProt ID	P30793
Immunogen	Recombinant protein of human GCH1

Background

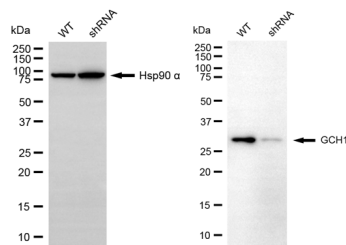
This gene encodes a member of the GTP cyclohydrolase family. The encoded protein is the first and rate-limiting enzyme in tetrahydrobiopterin (BH4) biosynthesis, catalyzing the conversion of GTP into 7,8-dihydroneopterin triphosphate. BH4 is an

essential cofactor required by aromatic amino acid hydroxylases as well as nitric oxide synthases. Mutations in this gene are associated with malignant hyperphenylalaninemia and dopa-responsive dystonia. Several alternatively spliced transcript variants encoding different isoforms have been described; however, not all variants give rise to a functional enzyme. [provided by RefSeq, Jul 2008]

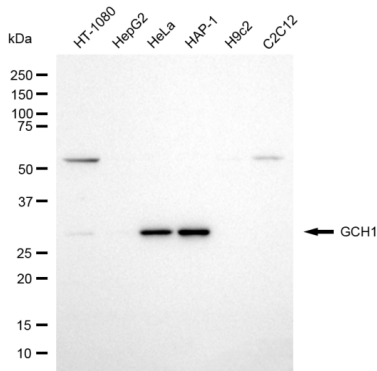
Research Area

Metabolism

Image Data



Western blotting analysis using GCH1 antibody (KVA00533). GCH1 expression in wild-type (WT) and GCH1 shRNA knockdown (KD) HeLa cells with 30 µg of total cell lysates. Hsp90 α serves as a loading control. The blot was incubated with GCH1 antibody (KVA00533, 1:2,500) and HRP-conjugated goat anti-mouse secondary antibody (APS0631, 1:10,000) respectively.



Western blotting analysis using GCH1 antibody (KVA00533). Total cell lysates (30 µg) from various cell lines were loaded and separated by SDS-PAGE. The blot was incubated with GCH1 antibody (KVA00533, 1:2,500) and HRP-conjugated goat anti-mouse secondary antibody (APS0631, 1:10,000) respectively.