



GALC (G-6): sc-518055

BACKGROUND

GALC (galactosylceramidase) is a lysosomal enzyme that hydrolyzes galactose ester bonds in various galactolipids, including galactosylceramide, galactosylsphingosine, lactosylceramide and monogalactosyldiglyceride. Galactolipids contain glucose and/or galactose, and are found in the brain and other nerve tissue, especially the myelin sheath. Galactosylceramide is a major lipid in myelin, kidney and epithelial cells of the small intestine and colon. Mutations in the GALC gene that compromise protein function correlate to Krabbe disease (globoid cell leukodystrophy, GLD). GLD is an autosomal recessive condition that affects approximately 1 in 150,000 infants and results in progressive destruction of the nervous system. The "twitcher" mouse is a model system for GLD; the genotype is a premature stop codon (W339X) in the galactosylceramidase (GALC) gene that abolishes enzymatic activity. Alternate transcriptional splice variants, encoding different isoforms, have been characterized.

REFERENCES

1. Kondo, Y., et al. 2005. Galactocerebroside-deficient oligodendrocytes maintain stable central myelin by exogenous replacement of the missing enzyme in mice. *Proc. Natl. Acad. Sci. USA* 102: 18670-18675.
2. Rafi, MA., et al. 2005. AAV-mediated expression of galactocerebroside symptoms and extended life span in murine models of globoid cell leukodystrophy. *Mol. Ther.* 11: 734-744.
3. Lin, D., et al. 2005. AAV2/5 vector expressing galactocerebroside ameliorates CNS disease in the murine model of globoid-cell leukodystrophy more efficiently than AAV2. *Mol. Ther.* 12: 422-430.
4. Meng, XL., et al. 2005. GALC transduction leads to morphological improvement of the twitcher oligodendrocytes *in vivo*. *Mol. Genet. Metab.* 84: 332-343.
5. Luzi, P., et al. 2005. Biochemical and pathological evaluation of long-lived mice with globoid cell leukodystrophy after bone marrow transplantation. *Mol. Genet. Metab.* 86: 150-159.

CHROMOSOMAL LOCATION

Genetic locus: GALC (human) mapping to 14q31.3.

SOURCE

GALC (G-6) is a mouse monoclonal antibody specific for an epitope mapping between amino acids 98-120 near the N-terminus of GALC 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.

GALC (G-6) is available conjugated to agarose (sc-518055 AC), 500 µg/0.25 ml agarose in 1 ml, for IP; to HRP (sc-518055 HRP), 200 µg/ml, for WB, IHC(P) and ELISA; to either phycoerythrin (sc-518055 PE), fluorescein (sc-518055 FITC), Alexa Fluor® 488 (sc-518055 AF488), Alexa Fluor® 546 (sc-518055 AF546), Alexa Fluor® 594 (sc-518055 AF594) or Alexa Fluor® 647 (sc-518055 AF647), 200 µg/ml, for WB (RGB), IF, IHC(P) and FCM; and to either Alexa Fluor® 680 (sc-518055 AF680) or Alexa Fluor® 790 (sc-518055 AF790), 200 µg/ml, for Near-Infrared (NIR) WB, IF and FCM.

APPLICATIONS

GALC (G-6) is recommended for detection of GALC isoforms 1 and 2 of 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).

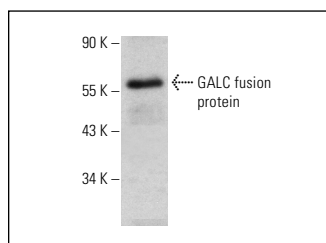
Suitable for use as control antibody for GALC siRNA (h): sc-60669, GALC shRNA Plasmid (h): sc-60669-SH and GALC shRNA (h) Lentiviral Particles: sc-60669-V.

Molecular Weight of GALC: 75 kDa.

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



GALC (G-6): sc-518055. Western blot analysis of human recombinant GALC fusion protein.

SELECT PRODUCT CITATIONS

1. Cuní-López, C., et al. 2024. Advanced patient-specific microglia cell models for pre-clinical studies in Alzheimer's disease. *J. Neuroinflammation* 21: 50.

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 for detailed protocols and support products.

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