

GALC (A-20): sc-49287

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

- 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.
- Rafi, M.A., 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.
- 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.
- Meng, X.L., et al. 2005. GALC transduction leads to morphological improvement of the twitcher oligodendrocytes *in vivo*. *Mol. Genet. Metab.* 84: 332-343.
- 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.
- Escobar, M.L., et al. 2005. Transplantation of umbilical-cord blood in babies with infantile Krabbe's disease. *N. Engl. J. Med.* 352: 2069-2081.
- Luddi, A., et al. 2005. Galactosylceramidase deficiency causes sperm abnormalities in the mouse model of globoid cell leukodystrophy. *Exp. Cell Res.* 304: 59-68.
- Beier, U.H., et al. 2005. Implications of galactocerebroside and galactosylcerebroside metabolism in cancer cells. *Int. J. Cancer* 115: 6-10.
- Pellegatta, S., et al. 2006. The therapeutic potential of neural stem/progenitor cells in murine globoid cell leukodystrophy is conditioned by macrophage/microglia activation. *Neurobiol. Dis.* 21: 314-323.

CHROMOSOMAL LOCATION

Genetic locus: GALC (human) mapping to 14q31.3; Galc (mouse) mapping to 12 E.

SOURCE

GALC (A-20) is an affinity purified goat polyclonal antibody raised against a peptide mapping within an internal region of GALC of human origin.

PRODUCT

Each vial contains 200 µg IgG in 1.0 ml of PBS with < 0.1% sodium azide and 0.1% gelatin.

Blocking peptide available for competition studies, sc-49287 P, (100 µg peptide in 0.5 ml PBS containing < 0.1% sodium azide and 0.2% BSA).

APPLICATIONS

GALC (A-20) is recommended for detection of GALC isoforms 1 and 2 of mouse, rat and human origin by Western Blotting (starting dilution 1:200, dilution range 1:100-1:1000), 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).

GALC (A-20) is also recommended for detection of GALC isoforms 1 and 2 in additional species, including equine, canine and bovine.

Suitable for use as control antibody for GALC siRNA (h): sc-60669, GALC siRNA (m): sc-60670, GALC shRNA Plasmid (h): sc-60669-SH, GALC shRNA Plasmid (m): sc-60670-SH, GALC shRNA (h) Lentiviral Particles: sc-60669-V and GALC shRNA (m) Lentiviral Particles: sc-60670-V.

Molecular Weight of GALC: 75 kDa.

RECOMMENDED SECONDARY REAGENTS

To ensure optimal results, the following support (secondary) reagents are recommended: 1) Western Blotting: use donkey anti-goat IgG-HRP: sc-2020 (dilution range: 1:2000-1:100,000) or Cruz Marker™ compatible donkey anti-goat IgG-HRP: sc-2033 (dilution range: 1:2000-1:5000), Cruz Marker™ Molecular Weight Standards: sc-2035, TBS Blotto A Blocking Reagent: sc-2333 and Western Blotting Luminol Reagent: sc-2048. 2) Immunofluorescence: use donkey anti-goat IgG-FITC: sc-2024 (dilution range: 1:100-1:400) or donkey anti-goat IgG-TR: sc-2783 (dilution range: 1:100-1:400) with UltraCruz™ Mounting Medium: sc-24941.

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 or our catalog for detailed protocols and support products.



Try **GALC (2D1): sc-293200**, our highly recommended monoclonal alternative to GALC (A-20).