

# GALC (H-300): sc-67352

## 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

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- 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.
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- 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.
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- Luddi, A., et al. 2005. Galactosylceramidase deficiency causes sperm abnormalities in the mouse model of globoid cell leukodystrophy. *Exp. Cell Res.* 304: 59-68.
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## CHROMOSOMAL LOCATION

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

## SOURCE

GALC (H-300) is a rabbit polyclonal antibody raised against amino acids 31-330 mapping near the N-terminus 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.

## APPLICATIONS

GALC (H-300) 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), 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), immunohistochemistry (including paraffin-embedded sections) (starting dilution 1:50, dilution range 1:50-1:500) and solid phase ELISA (starting dilution 1:30, dilution range 1:30-1:3000).

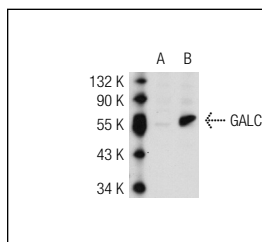
GALC (H-300) is also recommended for detection of GALC isoforms 1 and 2 in additional species, including equine, canine, bovine and porcine.

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.

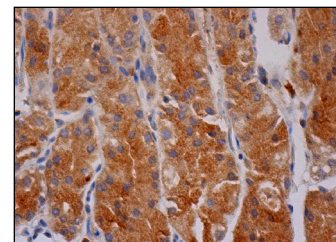
Molecular Weight of GALC: 75 kDa.

Positive Controls: GALC (h2): 293T Lysate: sc-170590.

## DATA



GALC (H-300): sc-67352. Western blot analysis of GALC expression in non-transfected: sc-117752 (A) and human GALC transfected: sc-170590 (B) 293T whole cell lysates.



GALC (H-300): sc-67352. Immunoperoxidase staining of formalin fixed, paraffin-embedded human upper stomach tissue showing cytoplasmic staining of glandular cells.

## 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.

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Satisfaction  
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Try **GALC (2D1): sc-293200**, our highly recommended monoclonal alternative to GALC (H-300).