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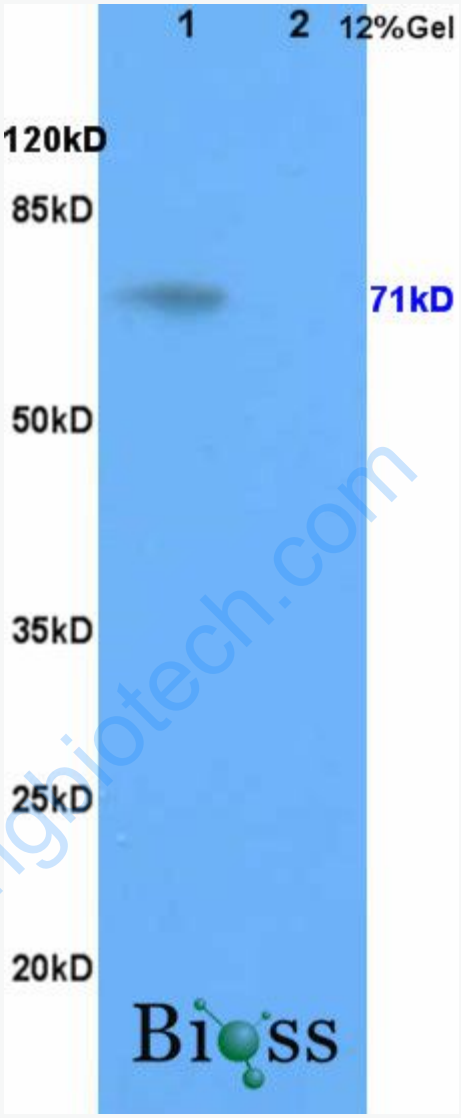
Rabbit Anti-Galactocerebrosidase antibody

SL4691R

Product Name:	Galactocerebrosidase
Chinese Name:	半乳糖脑苷脂酶抗体
Alias:	Gacy; Galc; Galactocerebroside beta galactosidase; Galactosylceramide beta-galactosidase; Galc; GALCERase; Twitcher; GALC_HUMAN; Galactocerebroside.
Organism Species:	Rabbit
Clonality:	Polyclonal
React Species:	Human,Mouse,Rat,Dog,Pig,Cow,Horse,Rabbit,Guinea Pig,
Applications:	WB=1:500-2000ELISA=1:500-1000 not yet tested in other applications. optimal dilutions/concentrations should be determined by the end user.
Molecular weight:	71kDa
Cellular localization:	cytoplasmic
Form:	Lyophilized or Liquid
Concentration:	1mg/ml
immunogen:	KLH conjugated synthetic peptide derived from human Galactocerebroside:301-400/685
Lsotype:	IgG
Purification:	affinity purified by Protein A
Storage Buffer:	0.01M TBS(pH7.4) with 1% BSA, 0.03% Proclin300 and 50% Glycerol.
Storage:	Store at -20 °C for one year. Avoid repeated freeze/thaw cycles. The lyophilized antibody is stable at room temperature for at least one month and for greater than a year when kept at -20°C. When reconstituted in sterile pH 7.4 0.01M PBS or diluent of antibody the antibody is stable for at least two weeks at 2-4 °C.
PubMed:	PubMed
Product Detail:	Galactosylceramidase hydrolyzes the galactose ester bonds of galactosylceramide, galactosylsphingosine, lactosylceramide, and monogalactosyldiglyceride. It is an enzyme with very low activity responsible for the lysosomal catabolism of galactosylceramide, a major lipid in myelin, kidney and epithelial cells of small intestine and colon. It shows highest level of activity in testes compared to brain, kidney, placenta and liver. It can also be found in urine. Defects in Galactosylceramidase are the

	cause of globoid cell leukodystrophy (GLD); also known as Krabbe disease. This autosomal recessive disorder results in the insufficient catabolism of several galactolipids that are important in the production of normal myelin. Clinically, the most frequent form is the infantile form. Most patients (90%) present before six months of age with irritability, spasticity, arrest of motor and mental development, and bouts of temperature elevation without infection. This is followed by myoclonic jerks of arms and legs, oposthotonus, hypertonic fits, and mental regression, which progresses to a severe decerebrate condition with no voluntary movements and death from respiratory infections or cerebral hyperpyrexia before 2 years of age. However, a significant number of cases with later onset, presenting with unexplained blindness, weakness and/or progressive motor, and sensory neuropathy that can progress to severe mental incapacity and death, have been identified. **Function:** Hydrolyzes the galactose ester bonds of galactosylceramide, galactosylsphingosine, lactosylceramide, and monogalactosyldiglyceride. Enzyme with very low activity responsible for the lysosomal catabolism of galactosylceramide, a major lipid in myelin, kidney and epithelial cells of small intestine and colon. **Subcellular Location:** Lysosomal. **Tissue Specificity:** Highest level of activity in testes compared to brain, kidney, placenta and liver. Can also be found in urine. **Similarity:** Belongs to the glycosyl hydrolase 59 family. **SWISS:** P54803 **Gene ID:** 2581 **Important Note:** This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.

Picture:



Sample:

Brain (Rat) Lysate at 40 ug

Liver (Rat) Lysate at 40 ug

Primary: Anti-Galactocerebroside (SL4691R) at 1/300 dilution

Secondary: HRP conjugated Goat-Anti-rabbit IgG (SL4691R) at 1/5000 dilution

Predicted band size: 71 kD

	Observed band size: 71 kD