

DESCRIPTION

Species Reactivity	Human
Specificity	Detects human β -Galactosidase-1/GLB1 in direct ELISAs.
Source	Monoclonal Mouse IgG _{2A} Clone # 810226
Purification	Protein A or G purified from hybridoma culture supernatant
Immunogen	Chinese hamster ovary cell line CHO-derived recombinant human β -Galactosidase-1/GLB1 Met1-Val677 Accession # P16278
Conjugate	Alexa Fluor 405 Excitation Wavelength: 405 nm Emission Wavelength: 421 nm
Formulation	Supplied 0.2mg/ml in 1X PBS with RDF1 and 0.09% Sodium Azide *Contains <0.1% Sodium Azide, which is not hazardous at this concentration according to GHS classifications. Refer to the Safety Data Sheet (SDS) for additional information and handling instructions.

APPLICATIONS

Please Note: Optimal dilutions should be determined by each laboratory for each application. [General Protocols](#) are available in the Technical Information section on our website.

Western Blot Optimal dilution of this antibody should be experimentally determined.

PREPARATION AND STORAGE

Shipping	The product is shipped with polar packs. Upon receipt, store it immediately at the temperature recommended below.
Stability & Storage	Protect from light. Do not freeze. 12 months from date of receipt, 2 to 8 °C as supplied

BACKGROUND

GLB1, a 60-76 kDa (predicted) glycoprotein, is a lysosomal β -galactosidase that hydrolyzes the terminal β -galactose from ganglioside and keratan sulfate. Defects in this gene are the causes of lysosomal storage diseases for GM1-gangliosidosis and Morquio B syndrome (also known as mucopolysaccharidosis IVB) (1, 2, 3). In GM1 gangliosidosis, GM1 ganglioside accumulates in the neurons of the central nervous system, because of the deficiency (0 $\pm$ 3% of normal) of lysosomal β -galactosidase activity. GM1 gangliosidosis demonstrates varying degrees of clinical severity but is invariably fatal, and children with the most common and severe form of GM1 gangliosidosis usually die within 3 years of birth. Morquio B syndrome patients are neurologically normal, but display severe skeletal dysostosis multiplex because of an accumulation of keratan sulfate (4). More than 100 mutations have been identified for GLB1, which result in different residual activities of the mutant enzymes and a spectrum of symptoms in the two related diseases (5). In lysosome, the mature β -galactosidase protein associates with cathepsin A and neuraminidase 1 to form the lysosomal multienzyme complex (6). An alternative splicing at the RNA level of GLB1 results a catalytically inactive β -galactosidase (also called elastin-binding protein) that plays an important role in vascular development (7).

PRODUCT SPECIFIC NOTICES

This product is provided under an agreement between Life Technologies Corporation and R&D Systems, Inc, and the manufacture, use, sale or import of this product is subject to one or more US patents and corresponding non-US equivalents, owned by Life Technologies Corporation and its affiliates. The purchase of this product conveys to the buyer the non-transferable right to use the purchased amount of the product and components of the product only in research conducted by the buyer (whether the buyer is an academic or for-profit entity). The sale of this product is expressly conditioned on the buyer not using the product or its components (1) in manufacturing; (2) to provide a service, information, or data to an unaffiliated third party for payment; (3) for therapeutic, diagnostic or prophylactic purposes; (4) to resell, sell, or otherwise transfer this product or its components to any third party, or for any other commercial purpose. Life Technologies Corporation will not assert a claim against the buyer of the infringement of the above patents based on the manufacture, use or sale of a commercial product developed in research by the buyer in which this product or its components was employed, provided that neither this product nor any of its components was used in the manufacture of such product. For information on purchasing a license to this product for purposes other than research, contact Life Technologies Corporation, Cell Analysis Business Unit, Business Development, 29851 Willow Creek Road, Eugene, OR 97402, Tel: (541) 465-8300. Fax: (541) 335-0354.