Human β-Galactosidase-1/GLB1
Alexa Fluor® 405-conjugated Antibody

Antigen Affinity-purified Polyclonal Sheep IgG
Catalog Number: AF6464V
100 µg

DESCRIPTION

Species Reactivity	Human
Specificity	Detects human β-Galactosidase-1/GLB1 in direct ELISAs and Western blots.
Source	Polyclonal Sheep IgG
Purification	Antigen Affinity-purified
Immunogen	Chinese hamster ovary cell line CHO-derived recombinant human β-Galactosidase-1/GLB1 Leu24-Val677 Accession # AAA51819
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±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

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