

DESCRIPTION

Species Reactivity	Human
Specificity	Detects human Lysosomal α -Glucosidase in direct ELISAs. Detects only 76Kda cleaved band.
Source	Monoclonal Rabbit IgG Clone # 2489B
Purification	Protein A or G purified from cell culture supernatant
Immunogen	Human embryonic kidney cell HEK293-derived human Lysosomal α -Glucosidase Ala70-Cys952 Accession # P10253
Conjugate	Alexa Fluor 594 Excitation Wavelength: 590 nm Emission Wavelength: 617 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.
Immunohistochemistry	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

Acid alpha-glucosidase (GAA) is an enzyme that is essential in the degradation of glycogen to glucose in the lysosome (1). Defects in GAA are the cause of glycogen storage disease II, also known as Pompe's disease, which is a rare autosomal recessive metabolic disorder that damages muscle and nerve cells throughout the body, primarily due to the accumulation of glycogen in the lysosome (2). Pompe disease occurs in babies, children, and adults who inherit a defective GAA gene and affects an estimated 5,000 to 10,000 people worldwide (3). Enzyme replacement therapy (ERT) is used to treat patients with this disease (4, 5).

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.