Human Lysosomal α-Glucosidase Alexa Fluor® 350-conjugated Antibody

Monoclonal Rabbit IgG Clone # 2489C
Catalog Number: FAB8329U
100 µg

DESCRIPTION

Species Reactivity	Human
Specificity	Detects human Lysosomal α-Glucosidase protein in direct ELISAs.
Source	Monoclonal Rabbit IgG Clone # 2489C
Purification	Protein A or G purified from cell culture supernatant
Immunogen	Human embryonic kidney cell, HEK293-derived human Lysosomal α-Glucosidase protein Ala70-Cys952 Accession # P10253
Conjugate	Alexa Fluor 350 Excitation Wavelength: 346 nm Emission Wavelength: 442 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).

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