

#### DESCRIPTION

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|---------------------------|-----------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------|
| <b>Species Reactivity</b> | Human                                                                                                                                                                                                                                                                       |
| <b>Specificity</b>        | Detects human Lysosomal $\alpha$ -Glucosidase in direct ELISAs. Detects only 76Kda cleaved band.                                                                                                                                                                            |
| <b>Source</b>             | Monoclonal Rabbit IgG Clone # 2489B                                                                                                                                                                                                                                         |
| <b>Purification</b>       | Protein A or G purified from cell culture supernatant                                                                                                                                                                                                                       |
| <b>Immunogen</b>          | Human embryonic kidney cell HEK293-derived human Lysosomal $\alpha$ -Glucosidase<br>Ala70-Cys952<br>Accession # P10253                                                                                                                                                      |
| <b>Conjugate</b>          | Alexa Fluor 700<br>Excitation Wavelength: 675-700 nm<br>Emission Wavelength: 723 nm                                                                                                                                                                                         |
| <b>Formulation</b>        | Supplied 0.2mg/ml in 1X PBS with RDF1 and 0.09% Sodium Azide<br><br>*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.

|                             |                                                                        |
|-----------------------------|------------------------------------------------------------------------|
| <b>Western Blot</b>         | Optimal dilution of this antibody should be experimentally determined. |
| <b>Immunohistochemistry</b> | Optimal dilution of this antibody should be experimentally determined. |

#### PREPARATION AND STORAGE

|                                |                                                                                                                   |
|--------------------------------|-------------------------------------------------------------------------------------------------------------------|
| <b>Shipping</b>                | The product is shipped with polar packs. Upon receipt, store it immediately at the temperature recommended below. |
| <b>Stability &amp; Storage</b> | 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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