

DESCRIPTION

Species Reactivity	Human
Specificity	Detects human alpha-Galactosidase A/GLA in direct ELISAs.
Source	Monoclonal Mouse IgG ₁ Clone # 1029734
Purification	Protein A or G purified from hybridoma culture supernatant
Immunogen	Chinese Hamster Ovary cell line CHO-derived human alpha-Galactosidase A/GLA protein Met1-Leu429 Accession # P06280
Conjugate	Alexa Fluor 647 Excitation Wavelength: 650 nm Emission Wavelength: 668 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.

Immunocytochemistry 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

Human α -Galactosidase A is a homodimeric glycoprotein that can release terminal α -galactosyl moieties from glycolipids and glycoproteins and catalyze the hydrolysis of melibiose into galactose and glucose (1). It is a lysosomal enzyme and is responsible for degradation of glycolipid globotriaosylceramide (Gb3) (Gal α 1-4Gal β 1-4Glc β -ceramide). Mutations in this gene cause Fabry disease, an X-linked hereditary lysosomal storage disease with the accumulation of Gb3 in the walls of small blood vessels, nerves, dorsal root ganglia, renal glomerular and tubular epithelial cells, and cardiomyocytes (2, 3). Inability to prevent the glycosphingolipid deposition can cause hypertension, strokes, heart attack and progressive renal failure (4). Current treatment for Fabry disease is enzyme replacement therapy using intravenously delivered recombinant α -Galactosidase A (5, 6).

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