Human α-Galactosidase A/GLA Antibody

Antigen Affinity-purified Polyclonal Sheep IgG
Catalog Number: AF6146

DESCRIPTION

Species Reactivity	Human
Specificity	Detects human α-Galactosidase A/GLA in direct ELISAs.
Source	Polyclonal Sheep IgG
Purification	Antigen Affinity-purified
Immunogen	Chinese hamster ovary cell line CHO-derived recombinant human α-Galactosidase A/GLA Leu32-Leu429 Accession # P06280
Formulation	Lyophilized from a 0.2 μm filtered solution in PBS with Trehalose. See Certificate of Analysis for details. *Small pack size (-SP) is supplied either lyophilized or as a 0.2 μm filtered solution in PBS.

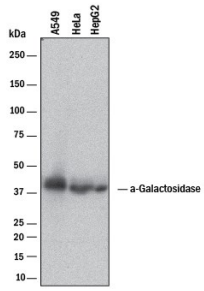
APPLICATIONS

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.*

	Recommended Concentration	Sample
Western Blot	2 µg/mL	See Below
Immunocytochemistry	5-15 µg/mL	See Below

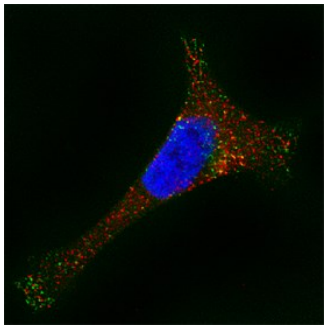
DATA

Western Blot



Detection of Human α-Galactosidase A/GLA by Western Blot. Western blot shows lysates of A549 human lung carcinoma cell line, HeLa human cervical epithelial carcinoma cell line, and HepG2 human hepatocellular carcinoma cell line. PVDF membrane was probed with 2 µg/mL of Sheep Anti-Human α-Galactosidase A/GLA Antigen Affinity-purified Polyclonal Antibody (Catalog # AF6146) followed by HRP-conjugated Anti-Sheep IgG Secondary Antibody (Catalog # HAF016). A specific band was detected for α-Galactosidase A/GLA at approximately 38 kDa (as indicated). This experiment was conducted under reducing conditions and using Immunoblot Buffer Group 1.

Immunocytochemistry



α-Galactosidase A/GLA in HeLa Human Cell Line. α-Galactosidase A/GLA was detected in immersion fixed HeLa human cervical epithelial carcinoma cell line using Sheep Anti-Human α-Galactosidase A/GLA Antigen Affinity-purified Polyclonal Antibody (Catalog # AF6146) at 15 µg/mL for 3 hours at room temperature. Cells were stained using the NorthernLights™ 557-conjugated Anti-Sheep IgG Secondary Antibody (red; Catalog # NL010). LAMP1 was also detected using Mouse Anti-Human LAMP1 Monoclonal Antibody (Catalog # MAB4800). Cells were stained using the NorthernLights™ 493-conjugated Anti-Mouse IgG Secondary Antibody (green; Catalog # NL009). Cells were counterstained with DAPI (blue). Specific staining was localized to lysosomes. View our protocol for Fluorescent ICC Staining of Cells on Coverslips.

PREPARATION AND STORAGE

Reconstitution	Sterile PBS to a final concentration of 0.2 mg/mL.
Shipping	The product is shipped at ambient temperature. Upon receipt, store it immediately at the temperature recommended below. *Small pack size (-SP) is shipped with polar packs. Upon receipt, store it immediately at -20 to -70 °C
Stability & Storage	**Use a manual defrost freezer and avoid repeated freeze-thaw cycles.** • 12 months from date of receipt, -20 to -70 °C as supplied. • 1 month, 2 to 8 °C under sterile conditions after reconstitution. • 6 months, -20 to -70 °C under sterile conditions after reconstitution.

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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) (Galα1-4Galβ1-4Glcβ-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).

References:

1. Ioannou, Y.A. *et al.* (1998) Biochem. J. **332**:789.
2. Koide, T. *et al.* (1990) FEBS Lett. **259**:353.
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4. Germain, D.P. (2002) Expert. Opin. Investig. Drugs. **11**:1467.
5. Barngrover, D. (2003) J. Biotechnol. **95**:280.
6. Mignani, R. and Cagnoli, L. (2004) J. Nephrol. **17**:354.

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