abcam

Product datasheet

Anti-GAA antibody ab55049

1 Image

Overview

Product name	Anti-GAA antibody
Description	Mouse monoclonal to GAA
Tested applications	**Suitable for:** WB
Species reactivity	**Reacts with:** Recombinant fragment **Predicted to work with:** Human
Immunogen	Recombinant fragment, corresponding to amino acids 851-953 of Human GAA

Properties

Form	Liquid
Storage instructions	Shipped at 4°C. Upon delivery aliquot and store at -20°C or -80°C. Avoid repeated freeze / thaw cycles.
Storage buffer	Preservative: None PBS, pH 7.2
Purity	Protein G purified
Clonality	Monoclonal
Isotype	IgG1
Light chain type	kappa

Applications

Our Abpromise guarantee covers the use of **ab55049** in the following tested applications.

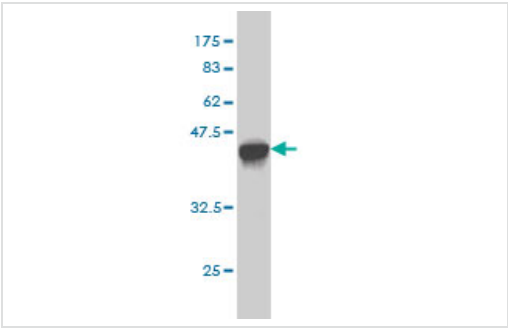
The application notes include recommended starting dilutions; optimal dilutions/concentrations should be determined by the end user.

Application	Abreviews	Notes
WB		Use a concentration of 1 - 5 µg/ml. This antibody has only been tested in WB against the recombinant fragment used as immunogen. We have no data on the detection of endogenous protein.

Target

Function	Essential for the degradation of glygogen to glucose in lysosomes.
Involvement in disease	Defects in GAA are the cause of glycogen storage disease type 2 (GSD2) [MIM:232300]; also called acid alpha-glucosidase (GAA) deficiency or acid maltase deficiency (AMD). GSD2 is a metabolic disorder with a broad clinical spectrum. The severe infantile form, or Pompe disease, presents at birth with massive accumulation of glycogen in muscle, heart and liver. Cardiomyopathy and muscular hypotonia are the cardinal features of this form whose life expectancy is less than two years. The juvenile and adult forms present as limb-girdle muscular dystrophy beginning in the lower limbs. Final outcome depends on respiratory muscle failure. Patients with the adult form can be free of clinical symptoms for most of their life but finally develop a slowly progressive myopathy.
Sequence similarities	Belongs to the glycosyl hydrolase 31 family. Contains 1 P-type (trefoil) domain.
Post-translational modifications	The different forms of acid glucosidase are obtained by proteolytic processing. Phosphorylation of mannose residues ensures efficient transport of the enzyme to the lysosomes via the mannose 6-phosphate receptor.
Cellular localization	Lysosome. Lysosome membrane.

Images



Western blot - GAA antibody (ab55049)

Western blot against tagged recombinant protein immunogen using ab55049 GAA antibody at 1ug/ml. Predicted band size of immunogen is 37 kDa.

This antibody has only been tested in WB against the recombinant fragment used as immunogen. We have no data on the detection of endogenous protein.

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