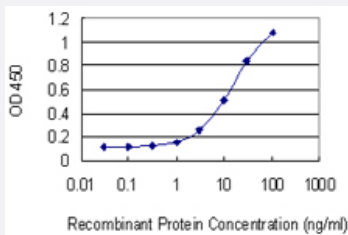


GAA monoclonal antibody (M01), clone 3C6

Catalog # H00002548-M01

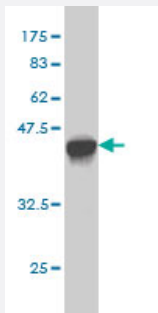
Size 100 ug

Applications



Sandwich ELISA (Recombinant protein)

Detection limit for recombinant GST tagged GAA is 0.3 ng/ml as a capture antibody.



Western Blot detection against Immunogen (36.96 KDa) .

Specification

Product Description	Mouse monoclonal antibody raised against a partial recombinant GAA.
Immunogen	GAA (AAH40431, 851 a.a. ~ 952 a.a) partial recombinant protein with GST tag. MW of the GST tag alone is 26 KDa.
Sequence	GEARGELFWDDGESLEVLERGAYTQVIFLARNTIVNELVRVTSEGAGLQLQKVTVLGVATAPQQ VLSNGVPVSNFTYSPDTKVLDCVSLLMGEQFLVSWC
Host	Mouse
Reactivity	Human
Isotype	IgG1 Kappa

Quality Control Testing

Antibody Reactive Against Recombinant Protein.
Western Blot detection against Immunogen (36.96 KDa) .

Storage Buffer

In 1x PBS, pH 7.4

Storage Instruction

Store at -20°C or lower. Aliquot to avoid repeated freezing and thawing.

Applications

- Western Blot (Recombinant protein)

[Protocol Download](#)

- Sandwich ELISA (Recombinant protein)

Detection limit for recombinant GST tagged GAA is 0.3 ng/ml as a capture antibody.

[Protocol Download](#)

- ELISA

Gene Info — GAA

Entrez GeneID

[2548](#)

GeneBank Accession#

[BC040431](#)

Protein Accession#

[AAH40431](#)

Gene Name

GAA

Gene Alias

LYAG

Gene Description

glucosidase, alpha; acid

Omim ID

[232300](#) [606800](#)

Gene Ontology

[Hyperlink](#)

Gene Summary

This gene encodes acid alpha-glucosidase, which is essential for the degradation of glycogen to glucose in lysosomes. Different forms of acid alpha-glucosidase are obtained by proteolytic processing. Defects in this gene are the cause of glycogen storage disease II, also known as Pompe's disease, which is an autosomal recessive disorder with a broad clinical spectrum. Three transcript variants encoding the same protein have been found for this gene. [provided by RefSeq

Other Designations

acid alpha-glucosidase|acid maltase|alpha-glucosidase|glycogen storage disease type II|lysosomal alpha-glucosidase

Publication Reference

- [Discovery of a novel noniminosugar acid \$\alpha\$ glucosidase chaperone series.](#)

Xiao J, Westbroek W, Motabar O, Lea WA, Hu X, Velayati A, Zheng W, Southall N, Gustafson AM, Goldin E, Sidransky E, Liu K, Simeonov A, Tamargo RJ, Ribes A, Matalonga L, Ferrer M, Marugan JJ.

Journal of Medicinal Chemistry 2012 Sep; 55(17):7546.

Application: WB, Human, Human fibroblasts, HEK 293

- [Glycosylation-related gene expression is linked to differentiation status in glioblastomas undifferentiated cells.](#)

Cheray M, Petit D, Forestier L, Karayan-Tapon L, Maftah A, Jauberteau MO, Battu S, Gallet FP, Lalloue F.

Cancer Letters 2011 Dec; 312(1):24.

Application: WB-Ce, Human, U87 MG, U251 cells

Pathway

- [Galactose metabolism](#)
- [Lysosome](#)
- [Metabolic pathways](#)
- [Starch and sucrose metabolism](#)

Disease

- [Cardiovascular Diseases](#)
- [Diabetes Mellitus](#)
- [Edema](#)
- [Genetic Predisposition to Disease](#)
- [Glycogen Storage Disease Type II](#)
- [Heart Diseases](#)