

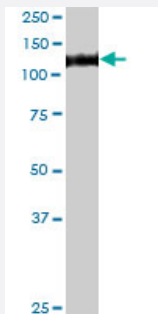
MaxPab®

GAA purified MaxPab rabbit polyclonal antibody (D01P)

Catalog # H00002548-D01P

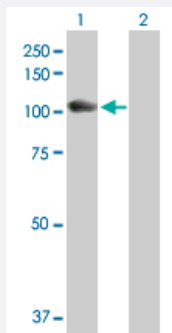
Size 100 ug

Applications



Western Blot (Tissue lysate)

GAA MaxPab rabbit polyclonal antibody. Western Blot analysis of GAA expression in mouse intestine.



Western Blot (Transfected lysate)

Western Blot analysis of GAA expression in transfected 293T cell line ([H00002548-T01](#)) by GAA MaxPab polyclonal antibody.

Lane 1: GAA transfected lysate(105.30 KDa).

Lane 2: Non-transfected lysate.

Specification

Product Description

Rabbit polyclonal antibody raised against a full-length human GAA protein.

Immunogen

GAA (P10253, 1 a.a. ~ 952 a.a) full-length human protein.

Sequence

MGVRHPPCSHRLLAVALVSLATAALLGHILLHDFLLVPRELSGSSPVLEETHPAHQQGASRPGP
RDAQAHPRPRAVPTQCDVPPNSRFDCAIDKAITQECCARGCCYIPAKQGLQGAQMGPWC
FFPPSYPSYKLENLSSSEMGYTATLTRTPTFFPKDILTRLVDMMETENRLHFTIKDPANRRYEVP
LETPRVHSRAPSPLYSVEFSEEPFGVIVHRQLDGRVLLNTTVAPLFFADQFLQLSTSLPSQYITGLA
EHLSPMLSTSWTRITLWNRDLAPTPGANLYGSHPFYLALEDGGSAGHGVLLNSNAMDVVLQPS
ALSWRSTGGILDVYIFLGPEPKSVVQQYLDVVGYPFMPYWGFGHLCRWGYSSTAITRQVVENM
TRAHFPLDVQWNLDDYMDSRDFTFNKDGFRDFPAMVQELHQGGRRYMMIVDPAISSSGPAGS
YRPYDEGLRRGVFITNETGQPLIGKVWPGSTAFPDFTNPTALAWWEDMVAEFHDQVPFDGMWID
MNEPSNFIRGSEDGCPNNELENPPYVPGVVGGTLQAATICASSHQFLSTHYNLHNLGLTEAIA
RALVKARGTRPFVISRSTFAGHGRYAGHWTGDVWSSWEQLASSVPEILQFNLLGVPLVGADVCG
FLGNTSEELCVRWTLQGAFFPFMRNHNLSLSPQEPYSFSEPAQQAMRKALTRYALLPHLYTLF
HQAHVAGETVARPLFLEFPKDSSTWTVDHQLLWGEALLITPVLQAGKAQVETGYFPLGTWYDLQTV
PIEALGSLPPPPAAPREPAIHSEGQWVTLPAPLDTINVHLRAGYIPLQGPGLTTESRQQPMALAV
ALTKGGEARGELFWDDGESLEVLERGAYTQVIFLARNTIVNELVRVTSEGAGLQLQKVTVLGVA
TAPQQVLSNGVPVSNFTYSPDTKVLDICVSLLMGEQFLVSWC

Host

Rabbit

Reactivity

Human, Mouse

Quality Control Testing

Antibody reactive against mammalian transfected lysate.

Storage Buffer

In 1x PBS, pH 7.4

Storage Instruction

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

Applications

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[Protocol Download](#)

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Western Blot analysis of GAA expression in transfected 293T cell line ([H00002548-T01](#)) by GAA MaxPab polyclonal antibody.

Lane 1: GAA transfected lysate(105.30 KDa).

Lane 2: Non-transfected lysate.

[Protocol Download](#)

Gene Info — GAA

Entrez GeneID

[2548](#)

GeneBank Accession#

[NM_000152.2](#)

Protein Accession#	P10253
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

- [Moss-Derived Human Recombinant GAA Provides an Optimized Enzyme Uptake in Differentiated Human Muscle Cells of Pompe Disease.](#)

Hintze S, Limmer S, Dabrowska-Schlepp P, Berg B, Krieghoff N, Busch A, Schaaf A, Meinke P, Schoser B.

International Journal of Molecular Sciences 2020 Apr; 21(7):2642.

Application: WB-Re, Recombinant protein

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](#)