GAA Rabbit mAb

Catalog No.: A19234 Recombinant



Basic Information

Observed MW

76kDa/105kDa

Calculated MW

105kDa

Category

Primary antibody

Applications

ELISA,WB

Cross-Reactivity

Human

CloneNo number

ARC2392

Background

This gene encodes lysosomal alpha-glucosidase, which is essential for the degradation of glycogen to glucose in lysosomes. The encoded preproprotein is proteolytically processed to generate multiple intermediate forms and the mature form of the enzyme. 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. Alternative splicing results in multiple transcript variants.

Recommended Dilutions

WB

1:500 - 1:1000

Immunogen Information

Gene ID

Swiss Prot

2548

P10253

Immunogen

A synthetic peptide corresponding to a sequence within amino acids 100-200 of human GAA (P10253).

Synonyms

LYAG; GAA

Contact

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Product Information

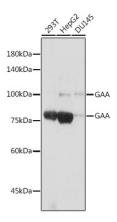
SourceIsotypePurificationRabbitIgGAffinity purification

Storage

Store at -20°C. Avoid freeze / thaw cycles.

Buffer: PBS with 0.02% sodium azide, 0.05% BSA, 50% glycerol, pH7.3.

Validation Data



Western blot analysis of various lysates using GAA Rabbit mAb (A19234) at 1:1000 dilution. Secondary antibody: HRP Goat Anti-Rabbit $\lg G$ (H+L) (AS014) at 1:10000 dilution.

Lysates/proteins: 25µg per lane.

Blocking buffer: 3% nonfat dry milk in TBST.

Detection: ECL Basic Kit (RM00020).

Exposure time: 10s.