

Product datasheet for TA334653

GAA Rabbit Polyclonal Antibody

Product data:

Product Type: Primary Antibodies

Applications: IHC, WB

Recommended Dilution: WB

Reactivity: Human
Host: Rabbit

Isotype: IgG

Clonality: Polyclonal

Immunogen: The immunogen for anti-GAA antibody: synthetic peptide directed towards the N terminal of

human GAA. Synthetic peptide located within the following region: FGVIVRRQLDGRVLLNTTVAPLFFADQFLQLSTSLPSQYITGLAEHLSPL

Formulation: Liquid. Purified antibody supplied in 1x PBS buffer with 0.09% (w/v) sodium azide and 2%

sucrose.

Note that this product is shipped as lyophilized powder to China customers.

Purification: Affinity Purified
Conjugation: Unconjugated

Storage: Store at -20°C as received.

Stability: Stable for 12 months from date of receipt.

Predicted Protein Size: 98 kDa

Gene Name: glucosidase alpha, acid

Database Link: NP 000143

Entrez Gene 2548 Human

P10253



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Background:

GAA is 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. 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.

Synonyms: LYAG

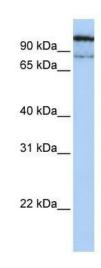
Note: Immunogen Sequence Homology: Dog: 100%; Pig: 100%; Rat: 100%; Human: 100%; Mouse:

100%; Bovine: 100%; Guinea pig: 100%; Horse: 93%; Zebrafish: 83%; Rabbit: 79%

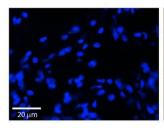
Protein Families: Druggable Genome, Transmembrane

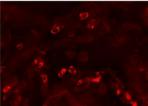
Protein Pathways: Galactose metabolism, Lysosome, Metabolic pathways, Starch and sucrose metabolism

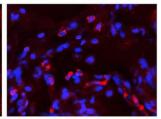
Product images:



WB Suggested Anti-GAA Antibody Titration: 0.2-1 ug/ml; ELISA Titer: 1: 62500; Positive Control: MCF7 cell lysate; GAA is supported by BioGPS gene expression data to be expressed in MCF7







Rabbit Anti-GAA Antibody; Formalin Fixed Paraffin Embedded Tissue: Human Lung Tissue; Observed Staining: Cytoplasmic in alveolar type I cells; Primary Antibody Concentration: 1: 100; Other Working Concentrations: 1/600; Secondary Antibody: Donkey anti-Rab