10320 Camino Santa Fe, Suite G
San Diego, CA 92121
Tel: 858.875.1900 Fax: 858.875.1999

# Anti-GAA Picoband Antibody

Catalog # ABO10192

## Product Information

| | |
|---|---|
| **Application** | WB, IHC-P |
| **Primary Accession** | P10253 |
| **Host** | Rabbit |
| **Reactivity** | Human, Rat |
| **Clonality** | Polyclonal |
| **Format** | Lyophilized |
| **Description** | Rabbit IgG polyclonal antibody for Lysosomal alpha-glucosidase(GAA) detection. Tested with WB, IHC-P in Human;Rat. |
| **Reconstitution** | Add 0.2ml of distilled water will yield a concentration of 500ug/ml. |

## Additional Information

| | |
|---|---|
| **Gene ID** | 2548 |
| **Other Names** | Lysosomal alpha-glucosidase, 3.2.1.20, Acid maltase, Aglucosidase alfa, 76 kDa lysosomal alpha-glucosidase, 70 kDa lysosomal alpha-glucosidase, GAA |
| **Calculated MW** | 105324 |
| **Application Details** | Immunohistochemistry(Paraffin-embedded Section), 0.5-1 □g/ml, Human, By Heat<br><br>Western blot, 0.1-0.5 □g/ml, Human, Rat |
| **Subcellular Localization** | Lysosome . Lysosome membrane . |
| **Source** | Eukaryota |
| **Protein Name** | Lysosomal alpha-glucosidase |
| **Contents** | Each vial contains 4mg Trehalose, 0.9mg NaCl, 0.2mg Na2HPO4, 0.05mg NaN3. |
| **Immunogen** | A synthetic peptide corresponding to a sequence in the middle region of human GAA (494-527aa TALAWWEDMVAEFHDQVPFDGMWIDMNEPSNFIR), different from the related mouse sequence by eight amino acids, and from the related rat sequence by six amino acids. |
| **Purification** | Immunogen affinity purified. |
| **Cross Reactivity** | No cross reactivity with other proteins |

| | |
|---|---|
| **Storage** | At -20˚C for one year. After r˚Constitution, at 4˚C for one month. It˚Can also be aliquotted and stored frozen at -20˚C for a longer time.Avoid repeated freezing and thawing. |

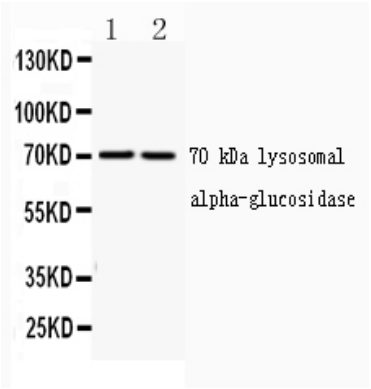
## Protein Information

| | |
|---|---|
| **Name** | GAA |
| **Function** | Essential for the degradation of glycogen in lysosomes (PubMed: 14695532, PubMed:18429042, PubMed:1856189, PubMed:7717400). Has highest activity on alpha-1,4-linked glycosidic linkages, but can also hydrolyze alpha-1,6-linked glucans (PubMed:29061980). |
| **Cellular Location** | Lysosome. Lysosome membrane |

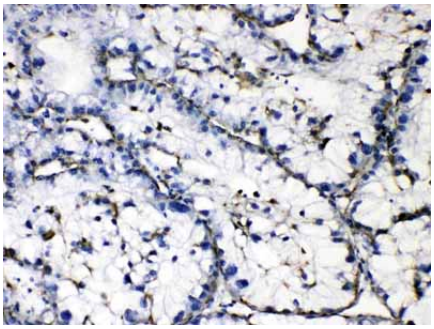
## Background

Lysosomal alpha-glucosidase is an enzyme that in humans is encoded by the GAA gene. 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.

## Images

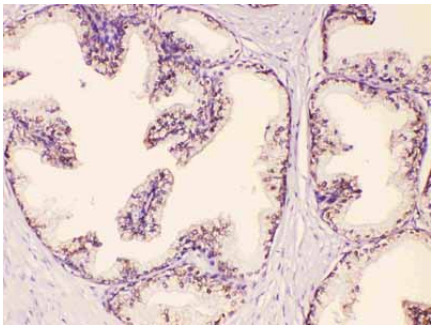



Western blot analysis of GAA expression in rat liver extract (lane 1) and A549 whole cell lysates (lane 2). GAA at 70KD was detected using rabbit anti- GAA Antigen Affinity purified polyclonal antibody (Catalog # ABO10192) at 0.5 □¼g/mL. The blot was developed using chemiluminescence (ECL) method .



GAA was detected in paraffin-embedded sections of human liver cancer tissues using rabbit anti- GAA Antigen Affinity purified polyclonal antibody (Catalog # ABO10192) at 1 □¼g/mL. The immunohistochemical section was developed using SABC method .

GAA was detected in paraffin-embedded sections of human prostatic cancer tissues using rabbit anti- GAA Antigen Affinity purified polyclonal antibody (Catalog # ABO10192) at 1 □¼g/mL. The immunohistochemical section was developed using SABC method .



Please note: All products are 'FOR RESEARCH USE ONLY. NOT FOR USE IN DIAGNOSTIC OR THERAPEUTIC PROCEDURES'.