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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 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.

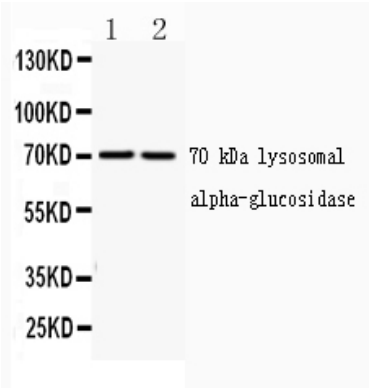
Protein Information

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

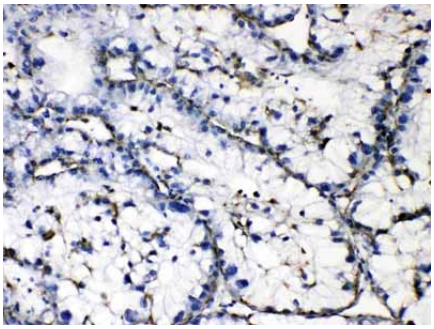
Background

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

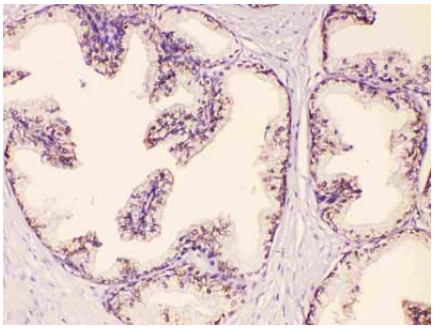


Western blot analysis of GAA expression in rat liver extract (lane 1) and A549 whole cell lysates (lane 2). GAA at 70KD was detected using rabbit anti- GAA Antigen Affinity purified polyclonal antibody (Catalog # ABO10192) at 0.5 □¼g/mL. The blot was developed using chemiluminescence (ECL) method .



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 .



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