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

GAA Antibody (N-term)

Affinity Purified Rabbit Polyclonal Antibody (Pab)
Catalog # AP12544a

Product Information

Application	IHC-P-Leica, IHC, WB, E
Primary Accession	P10253
Other Accession	NP_000143.2, NP_001073271.1
Reactivity	Human, Rat, Mouse
Host	Rabbit
Clonality	Polyclonal
Isotype	Rabbit IgG
Clone Names	RB18979
Calculated MW	105324
Antigen Region	174-203

Additional Information

Gene ID	2548
Other Names	Lysosomal alpha-glucosidase, Acid maltase, Aglucosidase alfa, 76 kDa lysosomal alpha-glucosidase, 70 kDa lysosomal alpha-glucosidase, GAA
Target/Specificity	This GAA antibody is generated from rabbits immunized with a KLH conjugated synthetic peptide between 174-203 amino acids from the N-terminal region of human GAA.
Dilution	IHC-P-Leica~~1:500 IHC~~1:100~500 WB~~1:1000 E~~Use at an assay dependent concentration.
Format	Purified polyclonal antibody supplied in PBS with 0.09% (W/V) sodium azide. This antibody is purified through a protein A column, followed by peptide affinity purification.
Storage	Maintain refrigerated at 2-8°C for up to 2 weeks. For long term storage store at -20°C in small aliquots to prevent freeze-thaw cycles.
Precautions	GAA Antibody (N-term) is for research use only and not for use in diagnostic or therapeutic procedures.

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

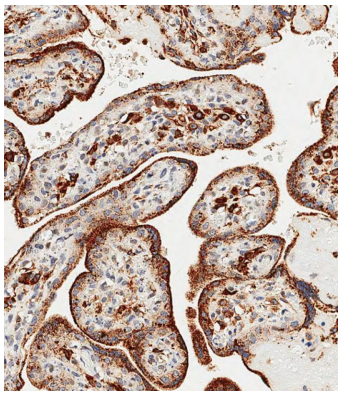
Background

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.

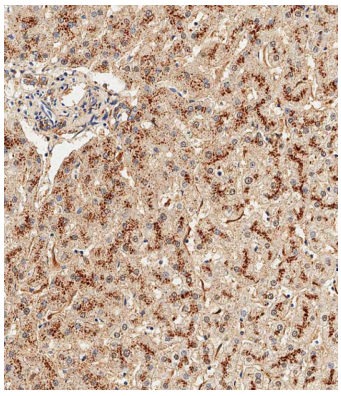
References

Bailey, S.D., et al. Diabetes Care 33(10):2250-2253(2010)
Labrousse, P., et al. Mol. Genet. Metab. 99(4):379-383(2010)
Talmud, P.J., et al. Am. J. Hum. Genet. 85(5):628-642(2009)
Aoyama, Y., et al. J. Hum. Genet. 54(11):681-686(2009)
Maimaiti, M., et al. J. Hum. Genet. 54(8):493-496(2009)

Images

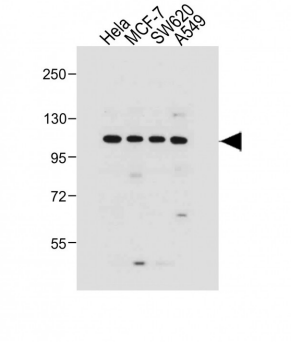


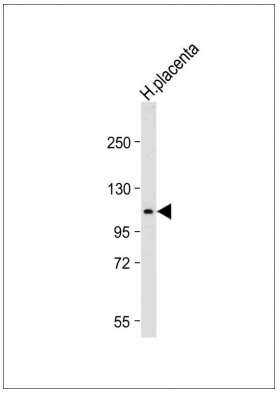
Immunohistochemical analysis of paraffin-embedded Human placenta tissue using AP12544a performed on the Leica® BOND RXm. Tissue was fixed with formaldehyde at room temperature, antigen retrieval was by heat mediation with a EDTA buffer (pH9. 0). Samples were incubated with primary antibody(1:500) for 1 hours at room temperature. A undiluted biotinylated CRF Anti-Polyvalent HRP Polymer antibody was used as the secondary antibody.



Immunohistochemical analysis of paraffin-embedded Human liver tissue using AP12544a performed on the Leica® BOND RXm. Tissue was fixed with formaldehyde at room temperature, antigen retrieval was by heat mediation with a EDTA buffer (pH9. 0). Samples were incubated with primary antibody(1:500) for 1 hours at room temperature. A undiluted biotinylated CRF Anti-Polyvalent HRP Polymer antibody was used as the secondary antibody.

All lanes : Anti-GAA Antibody (N-term) at 1:1000 dilution Lane 1: Hela whole cell lysate Lane 2: MCF-7 whole cell lysate Lane 3: SW620 whole cell lysate Lane 4: A549 whole cell lysate Lysates/proteins at 20 µg per lane. Secondary Goat Anti-Rabbit IgG, (H+L), Peroxidase conjugated at 1/10000 dilution. Predicted band size : 105 kDa Blocking/Dilution buffer: 5% NFDM/TBST.





Anti-GAA Antibody (N-term) at 1:1000 dilution + Human placenta tissue lysate Lysates/proteins at 20 µg per lane. Secondary Goat Anti-Rabbit IgG, (H+L), Peroxidase conjugated at 1/10000 dilution. Predicted band size : 105 kDa Blocking/Dilution buffer: 5% NFDM/TBST.