

**GAA Antibody (N-term)**  
**Affinity Purified Rabbit Polyclonal Antibody (Pab)**  
**Catalog # AP12544a**

**Specification**

---

**GAA Antibody (N-term) - Product Information**

Application	IHC-P-Leica, WB, IHC,E
Primary Accession	<a href="#">P10253</a>
Other Accession	<a href="#">NP_000143.2</a> , <a href="#">NP_001073271.1</a>
Reactivity	Human
Host	Rabbit
Clonality	Polyclonal
Isotype	Rabbit IgG
Antigen Region	174-203

**GAA Antibody (N-term) - 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

WB~~1:1000

IHC~~1:100~500

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.

**GAA Antibody (N-term) - 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**

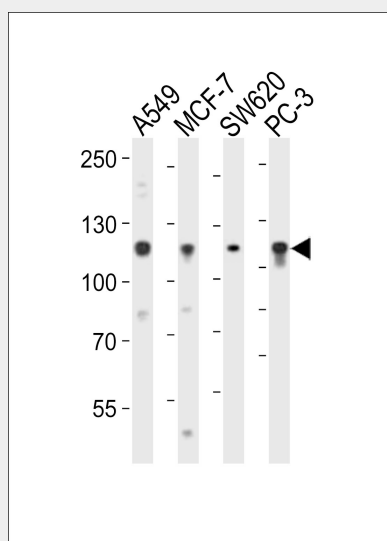
Lysosome. Lysosome membrane

**GAA Antibody (N-term) - Protocols**

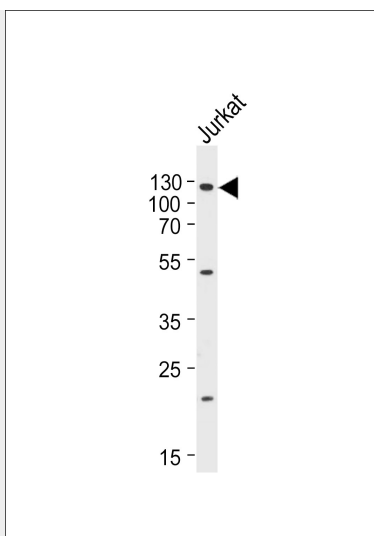
Provided below are standard protocols that you may find useful for product applications.

- [Western Blot](#)
- [Blocking Peptides](#)
- [Dot Blot](#)
- [Immunohistochemistry](#)
- [Immunofluorescence](#)
- [Immunoprecipitation](#)
- [Flow Cytometry](#)
- [Cell Culture](#)

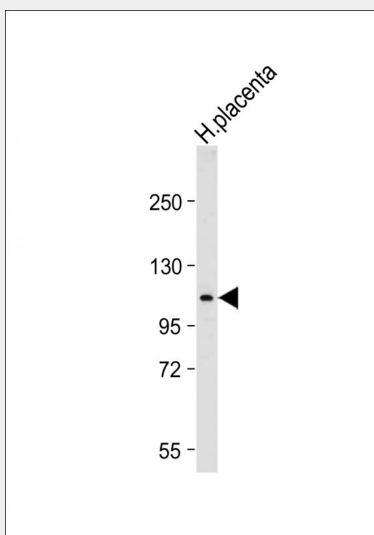
**GAA Antibody (N-term) - Images**



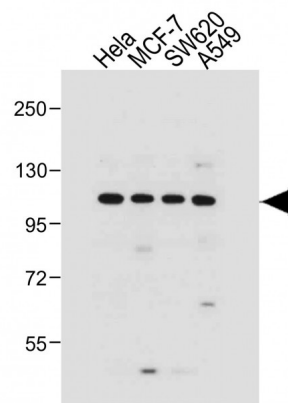
Western blot analysis of lysates from A549, MCF-7, SW620, PC-3 cell line (from left to right), using GAA Antibody (N-term)(Cat. #AP12544a). AP12544a was diluted at 1:1000 at each lane. A goat anti-rabbit IgG H&L(HRP) at 1:10000 dilution was used as the secondary antibody. Lysates at 20ug per lane.



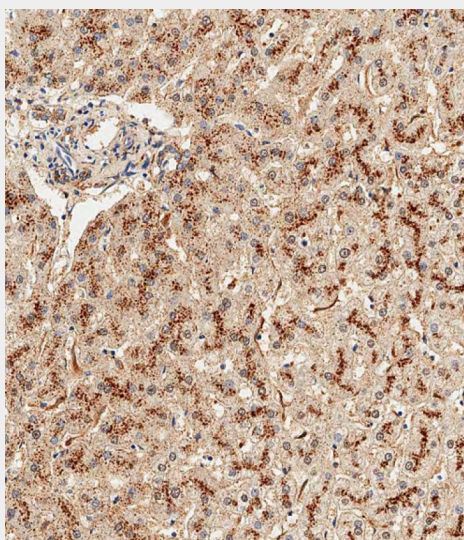
Western blot analysis of lysate from Jurkat cell line, using GAA Antibody (N-term)(Cat. #AP12544a). AP12544a was diluted at 1:2000. A goat anti-rabbit IgG H&L(HRP) at 1:10000 dilution was used as the secondary antibody. Lysate at 20ug.



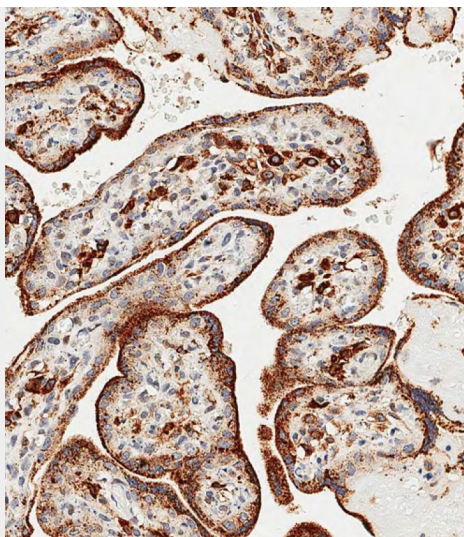
Anti-GAA Antibody (N-term) at 1:1000 dilution + Human placenta tissue lysate Lysates/proteins at 20 µg per lane. Secondary Goat Anti-Rabbit IgG, (H+L), Peroxidase conjugated at 1/10000 dilution. Predicted band size : 105 kDa Blocking/Dilution buffer: 5% NFDM/TBST.



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.



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.



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.

#### **GAA Antibody (N-term) - 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.

#### **GAA Antibody (N-term) - 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)