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

Specification

GAA Antibody (N-term) - Product Information

Application	**WB, IHC-P-Leica,E**
Primary Accession	P10253
Other Accession	NP_000143.2, NP_001073271.1
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
WB~~1:1000
IHC-P-Leica~~1:500

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:1856189,

PubMed:7717400, PubMed:14695532, PubMed:18429042). Has highest activity on alpha-1,4-linked glycosidic linkages, but can also hydrolyze alpha-1,6-linked glucans (PubMed:29061980).

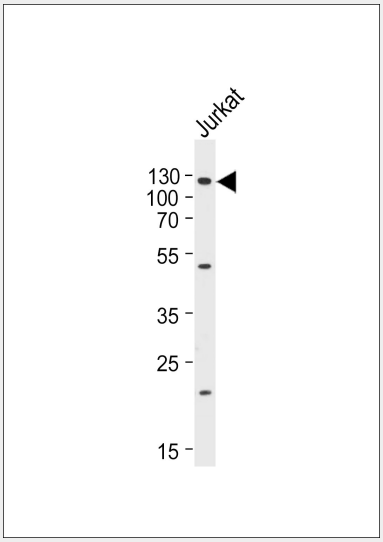
Cellular Location
Lysosome. Lysosome membrane

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 Cytomety
- Cell Culture

GAA Antibody (N-term) - Images



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.

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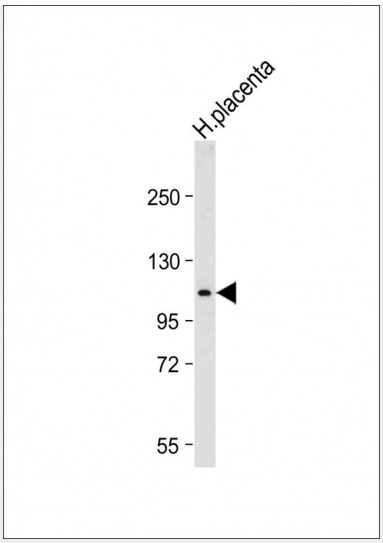
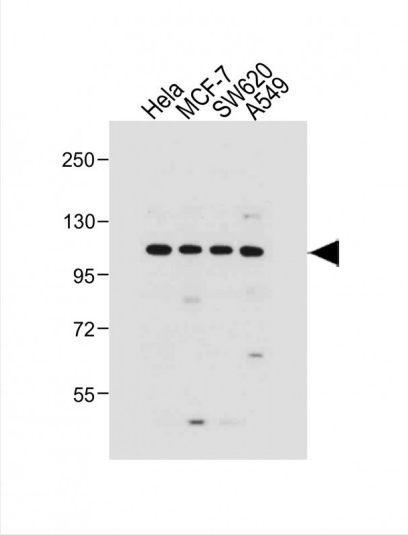


Anti-GAA Antibody (N-term) at 1:1000 dilution + Human placenta tissue lysate Lysates/proteins at 20 µg per lane. Secondary Goat Anti-Rabbit IgG, (H+L), Peroxidase conjugated at 1/10000 dilution. Predicted band size : 105 kDa Blocking/Dilution buffer: 5% NFDM/TBST.



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.

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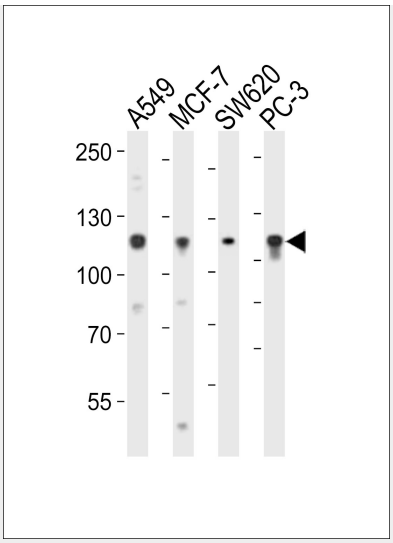
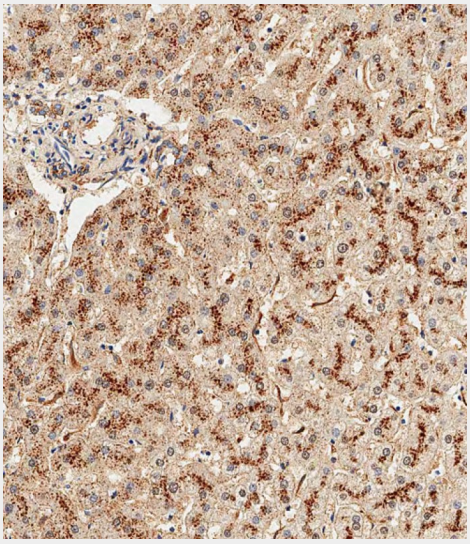


Western blot analysis of lysates from A549, MCF-7, SW620, PC-3 cell line (from left to right), using GAA Antibody (N-term)(Cat. #AP12544a). AP12544a was diluted at 1:1000 at each lane. A goat anti-rabbit IgG H&L(HRP) at 1:10000 dilution was used as the secondary antibody. Lysates at 20ug per lane.



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.

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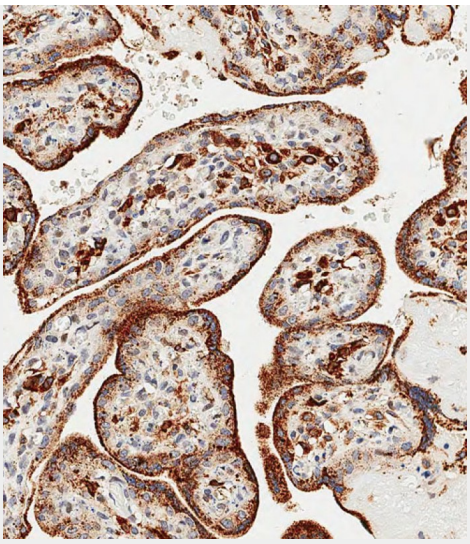

Immunohistochemical analysis of paraffin-embedded Human placenta tissue using AP12544a performed on the Leica® BOND RXm. Tissue was fixed with formaldehyde at room temperature, antigen retrieval was by heat mediation with a EDTA buffer (pH9. 0). Samples were incubated with primary antibody(1:500) for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