

Datasheet for ABIN7602983
anti-GAA antibody (Middle Region)



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Overview

Quantity:	100 µg
Target:	GAA
Binding Specificity:	Middle Region
Reactivity:	Human
Host:	Mouse
Clonality:	Monoclonal
Conjugate:	This GAA antibody is un-conjugated
Application:	Western Blotting (WB), Immunohistochemistry (IHC), Immunocytochemistry (ICC), Immunofluorescence (IF)

Product Details

Purpose:	Anti-GAA Antibody Picoband® (monoclonal, 2G7)
Immunogen:	A synthetic peptide corresponding to a sequence in the middle region of human GAA, different from the related mouse sequence by eight amino acids, and from the related rat sequence by six amino acids.
Clone:	2G7
Isotype:	IgG2b
Cross-Reactivity (Details):	No cross-reactivity with other proteins.
Characteristics:	Anti-GAA Antibody Picoband® (monoclonal, 2G7) (ABIN7602983). Tested in IF, IHC, ICC, WB applications. This antibody reacts with Human. The brand Picoband indicates this is a premium antibody that guarantees superior quality, high affinity, and strong signals with minimal

Product Details

background in Western blot applications. Only our best-performing antibodies are designated as Picoband, ensuring unmatched performance.

Purification: Immunogen affinity purified.

Target Details

Target: GAA

Alternative Name: GAA ([GAA Products](#))

Background: Synonyms: Lysosomal alpha-glucosidase,3.2.1.20
Tissue Specificity: Expressed on platelets and leukocytes and is primarily concentrated at the borders between endothelial cells. Expressed in human umbilical vein endothelial cells (HUVECs) (at protein level). Expressed on neutrophils (at protein level). Isoform Long predominates in all tissues examined. Isoform Delta12 is detected only in trachea. Isoform Delta14-15 is only detected in lung. Isoform Delta14 is detected in all tissues examined with the strongest expression in heart. Isoform Delta15 is expressed in brain, testis, ovary, cell surface of platelets, human umbilical vein endothelial cells (HUVECs), Jurkat T-cell leukemia, human erythroleukemia (HEL) and U-937 histiocytic lymphoma cell lines (at protein level).
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.

Molecular Weight: 110 kDa, 95 kDa, 76 kDa

Gene ID: 2548

UniProt: [P10253](#)

Pathways: [Cellular Glucan Metabolic Process](#)

Application Details

Application Notes: Western blot, 0.1-0.5 µg/mL, Human
Immunohistochemistry (Paraffin-embedded Section), 0.5-1 µg/mL, Human
Immunocytochemistry/Immunofluorescence, 2 µg/mL, Human

Application Details

1. "Entrez Gene: GAA glucosidase, alpha, acid (Pompe disease, glycogen storage disease type II)". 2. Donald J. Voet, Judith G. Voet, Charlotte W. Pratt (2008). "Additional Pathways in Carbohydrate Metabolism". Principles of Biochemistry, Third edition. Wiley. p. 538. 3. Reuser AJ, Kroos MA, Hermans MM, et al. (1995). "Glycogenosis type II (acid maltase deficiency)". Muscle Nerve. 3: S61-9.

Restrictions: For Research Use only

Handling

Format: Lyophilized

Reconstitution: Add 0.2 mL of distilled water will yield a concentration of 500 µg/mL.

Concentration: 500 µg/mL

Buffer: Each vial contains 4 mg Trehalose, 0.9 mg NaCl, 0.2 mg Na₂HPO₄, 0.01 mg Sodium azide.

Preservative: Sodium azide

Precaution of Use: This product contains Sodium azide: a POISONOUS AND HAZARDOUS SUBSTANCE which should be handled by trained staff only.

Storage: 4 °C, -20 °C

Storage Comment: Store at -20°C for one year from date of receipt. After reconstitution, at 4°C for one month. It can also be aliquotted and stored frozen at -20°C for six months. Avoid repeated freeze-thaw cycles.