abcam

Product datasheet

Recombinant Human GAA protein ab114893

画像数 1

製品の詳細

製品名	Recombinant Human GAA protein
精製度	>= 80 % Purified via GST Tag. Glutathione Sepharose
発現系	Wheat germ
アクセッション番号	P10253
タンパク質長	Full length protein
Animal free	No
由来	Recombinant
生物種	Human
配列	MGVRHPPCSHRLLAVCALVSLATAALLGHILLHDFLLVPREL SGSSPVLE ETHPAHQQGASRPGPRDAQAHPGRPRAVPTQCDVPPNSRFDC APDKAITQ EQCEARGCCYIPAKQGLQGAQMGQPWCFFPPSYPSYKLENLS SSEMGYTA TLTRTTPTFFPKDILTLRLDVMMETENRLHFTIKDPANRRYE VPLETPRV HSRAPSPLYSVEFSEEPFGVIVHRQLDGRVLLNTTVAPLFFA DQFLQLST SLPSQYITGLAEHLSPLMLSTSWTRITLWNRDLAPTPGANLY GSHPFYLA LEDGGSAHGVFLLNSNAMDVVLQPSPALSWRSTGGILDVYIF LGPEPKSV VQQYLDVVGYPFMPPYWGLGFHLCRWGYSSTAITRQVVENMT RAHFPLDV QWNDLDYMDSRRDFTFNKDGFRDFPAMVQELHQGGRRYMMIV DPAISSSG PAGSYRPYDEGLRRGVFITNETGQPLIGKVWPGSTAFPDFTN PTALAWWE DMVAEFHDQVPFDGMWIDMNEPSNFIRGSEDGCPNNELENPP YVPGVVGG TLQAATICASSHQFLSTHYNLHNLYGLTEAIASHRALVKARG TRPFVISR STFAGHGRYAGHWTGDVWSSWEQLASSVPEILQFNLLGVPLV

GADVCGFL
GNTSEELCVRWTQLGAFYPFMRNHNSLLSLPQEPYSFSEPAQ
QAMRKALT
LRYALLPHLYTLFHQAHVAGETVARPLFLEFPKDSSTWTVDH
QLLWGEAL
LITPVLQAGKAEVTGYFPLGTWYDLQTVPIEALGSLPPPPAA
PREPAIHS
EGQWVTLPAPLDTINVHLRAGYIIPLQGPGLTTTESRQQPMA
LAVALTKG
GEARGELFWDDGESLEVLERGAYTQVIFLARNNTIVNELVRV
TSEGAGLQ
LQKVTVLGVATAPQQVLSNGVPVSNFTYSPDTKVLDICVSLL
MGEQFLVS WC

予測される分子量 131 kDa including tags

領域 1 to 952

特性

Our **Abpromise guarantee** covers the use of **ab114893** in the following tested applications.

The application notes include recommended starting dilutions; optimal dilutions/concentrations should be determined by the end user.

アプリケーション ELISA

SDS-PAGE

Western blot

製品の状態 Liquid

前処理および保存

保存方法および安定性 Shipped on dry ice. Upon delivery aliquot and store at -80ºC. Avoid freeze / thaw cycles.

pH: 8.00
Constituents: 0.3% Glutathione, 0.79% Tris HCl

関連情報

機能 Essential for the degradation of glygogen to glucose in lysosomes.

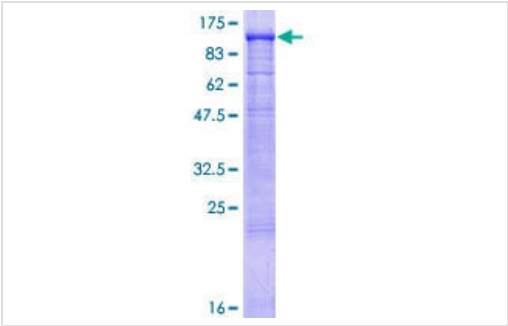
関連疾患 Defects in GAA are the cause of glycogen storage disease type 2 (GSD2) [MIM:232300]; also called acid alpha-glucosidase (GAA) deficiency or acid maltase deficiency (AMD). GSD2 is a metabolic disorder with a broad clinical spectrum. The severe infantile form, or Pompe disease, presents at birth with massive accumulation of glycogen in muscle, heart and liver. Cardiomyopathy and muscular hypotonia are the cardinal features of this form whose life expectancy is less than two years. The juvenile and adult forms present as limb-girdle muscular dystrophy beginning in the lower limbs. Final outcome depends on respiratory muscle failure. Patients with the adult form can be free of clinical symptoms for most of their life but finally develop a slowly progressive myopathy.

配列類似性 Belongs to the glycosyl hydrolase 31 family.
Contains 1 P-type (trefoil) domain.

翻訳後修飾 The different forms of acid glucosidase are obtained by proteolytic processing.

	Phosphorylation of mannose residues ensures efficient transport of the enzyme to the lysosomes via the mannose 6-phosphate receptor.
細胞内局在	Lysosome. Lysosome membrane.

画像



SDS-PAGE - Recombinant Human GAA protein (ab114893)

12.5% SDS-PAGE showing ab114893 at approximately 130.79kDa.
Stained with Coomassie Blue.

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- Response to your inquiry within 24 hours
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