

# Recombinant Human GBE1

**Cat. No.** GBE1-28988TH    **Lot. No.** (See product label)

## SPECIFICATION

<b>Product Overview</b>	Recombinant full length Human GBE1 with a N terminal proprietary tag: predicted molecular weight 103.29 kDa.
<b>Species</b>	Human
<b>Source</b>	Wheat Germ
<b>ProteinLength</b>	702 amino acids
<b>Description</b>	The protein encoded by this gene is a glycogen branching enzyme that catalyzes the transfer of alpha-1,4-linked glucosyl units from the outer end of a glycogen chain to an alpha-1,6 position on the same or a neighboring glycogen chain. Branching of the chains is essential to increase the solubility of the glycogen molecule and, consequently, in reducing the osmotic pressure within cells. Highest level of this enzyme are found in liver and muscle. Mutations in this gene are associated with glycogen storage disease IV (also known as Andersens disease).
<b>Molecular Weight</b>	103.290kDa inclusive of tags
<b>Tissue specificity</b>	Highest levels found in liver and muscle.
<b>Form</b>	Liquid
<b>Purity</b>	Proprietary Purification

 Tel: 1-631-559-9269    1-516-512-3133

 Email: [info@creative-biomart.com](mailto:info@creative-biomart.com)     Fax: 1-631-938-8127

 45-1 Ramsey Road, Shirley, NY 11967, USA

<b>Storage buffer</b>	pH: 8.00 Constituents: 0.3% Glutathione, 0.79% Tris HCl
<b>Storage</b>	Shipped on dry ice. Upon delivery aliquot and store at -80oC. Avoid freeze / thaw cycles.
<b>Sequences of amino acids</b>	<p>MAAPMTPAARPEDYEAALNAALADVPELARLLEIDPYLKP YAVDFQRRYKQFSQILK          NIGENEGGIDKFSRGYESFGVHR CADGGLYCKEWAPGAEGVFLTGFNGWNPFS          YPYKKLDYG KWELYIPPKQNKSVLVPHGSKLKVVITSKSGEILYRISPW AKYVVREG          DNVNYDWIHWDPHEHSYEFKHSRPPKPRSLRIY ESHVGISSHEGKVASYKHFTCNVL          PRIKGLGYNCIQLMAI MEHAYYASFGYQITSFFAASSRYGSPEELQELVDTAHSMT II          VLLDVVHSHASKNSADGLNMFDTGTDSCYFHSGPRGTHD LWDSRLFAYSSWEVLR          LLSNIRWWLEEYRFDGFRFDGVT SMLYHHHGVGQGFSGDYSEYFGLQVDEDALTY          LMLANHLV HTLCPDSITIAEDVSGMPALCSPISQGGGGFDYRLAMAIP DKWIQLLKE          FKDEDWNMGDIVYTLNRRYLEKCIAYAESH DQALVGDKSLAFWLMDAEMYTNMS          VLTPTPVIDRGIQLH KMIRLITHGLGGEGYLNFMGNEFGHPWLDLDFPRKGNNESY          HYARRQFHLTDDDLLRYKFLNNFDRDMNRLEERYGWLAAP QAYVSEKHEGNKIIAF          ERAGLLFIFNFHPSKSYTDYRVGT ALPGKFKIVLDSDAAEYGGHQRLDHSTDFFFSEA          FEHNGRP YSLLVYIPSRVALILQNVDLPN</p>
<b>Sequence Similarities</b>	Belongs to the glycosyl hydrolase 13 family.
<b>GENE INFORMATION</b>	
<b>Gene Name</b>	GBE1 glucan (1,4-alpha-), branching enzyme 1 [ Homo sapiens ]
<b>Official Symbol</b>	GBE1
<b>Synonyms</b>	GBE1; glucan (1,4-alpha-), branching enzyme 1; 1,4-alpha-glucan-branching enzyme; Andersen disease; glycogen branching enzyme; glycogen storage disease type IV;

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<b>Gene ID</b>	2632
<b>mRNA Refseq</b>	NM_000158
<b>Protein Refseq</b>	NP_000149
<b>MIM</b>	607839
<b>Uniprot ID</b>	Q04446
<b>Chromosome Location</b>	3
<b>Pathway</b>	Glucose metabolism, organism-specific biosystem; Glycogen Metabolism, organism-specific biosystem; Glycogen synthesis, organism-specific biosystem; Metabolic pathways, organism-specific biosystem; Metabolism of carbohydrates, organism-specific biosystem;
<b>Function</b>	1,4-alpha-glucan branching enzyme activity; cation binding; hydrolase activity, hydrolyzing O-glycosyl compounds; transferase activity, transferring glycosyl groups;

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