Datasheet for ABIN1707697

anti-GBE1 antibody (AA 101-200) (Cy7)

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Overview

Quantity:	100 µL
Target:	GBE1
Binding Specificity:	AA 101-200
Reactivity:	Human
Host:	Rabbit
Clonality:	Polyclonal
Conjugate:	This GBE1 antibody is conjugated to Cy7
Application:	Western Blotting (WB), Immunofluorescence (Cultured Cells) (IF (cc)), Immunofluorescence (Paraffin-embedded Sections) (IF (p))

Product Details

Immunogen:	KLH conjugated synthetic peptide derived from human GBE1
Isotype:	IgG
Predicted Reactivity:	Human,Mouse,Rat,Rabbit
Purification:	Purified by Protein A.

Target Details

Target:	GBE1
Alternative Name:	GBE1 (GBE1 Products)
Background:	Synonyms: 1,4 alpha glucan branching enzyme, 4-alpha-glucan-branching enzyme, amylo 1,4 to

Target Details

	1,6 transglucosidase, amylo 1,4 to 1,6 transglycosylase, Andersen disease, Brancher enzyme, GBE 1, GBE, GBE1, gGlucan 1,4 alpha , branching enzyme 1, GLGB_HUMAN, Glucan 1,4 alpha branching enzyme, Glycogen branching enzyme, Glycogen storage disease type IV, Glycogen-branching enzyme, OTTHUMP00000213788, OTTHUMP00000213833. Background: GBE1 is a 702 amino acid protein that is expressed at high levels in muscle and liver and is involved in glycogen biosynthesis. Existing as a monomer, GBE1 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 a neighboring glycogen chain and, via this catalytic activity, plays an essential role in glycogen accumulation. Defects in the gene encoding GBE1 are the cause of glycogen storage disease type 4 (GSD4) and adult polyglucosan body disease (APBD), the first of which is a metabolic disorder that is associated with the accumulation of polysaccharides and is characterized by liver disease during childhood. Unlike GSD4, APBD is a late-onset disorder that affects the central and peripheral nervous systems and is characterized by cognitive impairment, pyramidal tetraparesis and peripheral neuropathy.
Gene ID:	2632
Pathways:	Cellular Glucan Metabolic Process

Application Details

Application Notes:	IF(IHC-P) 1:50-200
	IF(IHC-F) 1:50-200
	IF(ICC) 1:50-200

Restrictions:	For Research Use only

Handling

Format:	Liquid
Concentration:	1 µg/µL
Buffer:	Aqueous buffered solution containing 0.01M TBS (pH 7.4) with 1 % BSA, 0.03 % Proclin300 and 50 % Glycerol.
Preservative:	ProClin
Precaution of Use:	This product contains ProClin: a POISONOUS AND HAZARDOUS SUBSTANCE, which should be handled by trained staff only.
Storage:	-20 °C

Handling

Storage Comment:	Store at -20°C. Aliquot into multiple vials to avoid repeated freeze-thaw cycles.
Expiry Date:	12 months