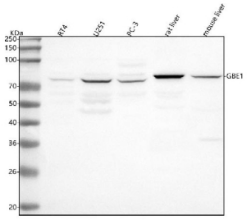
GBE1 Antibody / 1,4-alpha-glucan branching enzyme 1 (FY12899)

Catalog No.	Formulation	Size
FY12899	Adding 0.2 ml of distilled water will yield a concentration of 500 ug/ml	100 ug

Bulk quote request

Availability	1-2 days
Species Reactivity	Human, Mouse, Rat
Format	Lyophilized
Host	Rabbit
Clonality	Polyclonal (rabbit origin)
Isotype	Rabbit IgG
Purity	Immunogen affinity purified
Buffer	Each vial contains 4 mg Trehalose, 0.9 mg NaCl, 0.2 mg Na2HPO4.
UniProt	Q04446
Applications	Western Blot: 0.25-0.5ug/ml ELISA: 0.1-0.5ug/ml
Limitations	This GBE1 antibody is available for research use only.



Western blot analysis of GBE1 using anti-GBE1 antibody. Lane 1: human RT4 whole cell lysates, Lane 2: human U251 whole cell lysates, Lane 3: human PC-3 whole cell lysates, Lane 4: rat liver tissue lysates, Lane 5: mouse liver tissue lysates. After electrophoresis, proteins were transferred to a nitrocellulose membrane at 150 mA for 50-90 minutes. Blocked the membrane with 5% non-fat milk/TBS for 1.5 hour at RT. The membrane was incubated with rabbit anti-GBE1 antibody at 0.5 ug/ml overnight at 4oC, then washed with TBS-0.1%Tween 3 times with 5 minutes each and probed with a goat anti-rabbit IgG-HRP secondary antibody at a dilution of 1:5000 for 1.5 hour at RT. The signal was developed using enhanced chemiluminescent. A specific band was detected for GBE1 at approximately 80 kDa. The expected molecular weight of GBE1 is ~80 kDa.

Description

GBE1 antibody detects 1,4-alpha-glucan branching enzyme 1, an essential enzyme in glycogen biosynthesis responsible for introducing alpha-1,6 glycosidic branches into linear glucan chains. Encoded by the GBE1 gene on chromosome 3p12.3, this enzyme catalyzes the formation of glycogen's highly branched structure, which enhances solubility and allows rapid synthesis and mobilization of glucose. GBE1 is a crucial metabolic regulator in liver, skeletal muscle, and

other tissues involved in energy storage and glucose homeostasis.

Structurally, GBE1 is a 702-amino-acid cytosolic enzyme of approximately 80 kilodaltons that belongs to the glycoside hydrolase family 13. It contains a central catalytic domain with the conserved Asp-Glu-Asp catalytic triad and carbohydrate-binding modules that enable glucan chain recognition and rearrangement. By transferring short alpha-1,4-linked oligosaccharide chains to alpha-1,6 positions, GBE1 generates the branch points that define glycogen's compact and accessible configuration.

The GBE1 antibody is widely used in glycogen metabolism, endocrinology, and neuromuscular research to study carbohydrate storage, enzymatic regulation, and metabolic disease mechanisms. Western blot analysis detects an 80 kilodalton band corresponding to GBE1, while immunofluorescence shows diffuse cytoplasmic staining, particularly in hepatocytes and myocytes. This antibody supports investigations of glycogen biosynthesis and the enzymatic defects underlying glycogen storage disorders.

Mutations in GBE1 cause Glycogen Storage Disease Type IV (Andersen disease) and its non-lethal variant, adult polyglucosan body disease (APBD). These disorders are characterized by abnormal glycogen accumulation and progressive neuromuscular or hepatic dysfunction. Reduced GBE1 activity leads to poorly branched glycogen with decreased solubility and impaired mobilization. The GBE1 antibody provides a reliable tool for studying enzyme expression and activity in models of metabolic disease, helping to elucidate molecular mechanisms that govern glycogen structure and energy regulation. NSJ Bioreagents validates this antibody for its applications, ensuring high specificity and reproducibility in metabolic research.

Application Notes

Optimal dilution of the GBE1 antibody should be determined by the researcher.

Immunogen

E.coli-derived human GBE1 recombinant protein (Position: H79-R515) was used as the immunogen for the GBE1 antibody.

Storage

After reconstitution, the GBE1 antibody can be stored for up to one month at 4oC. For long-term, aliquot and store at -20oC. Avoid repeated freezing and thawing.

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