

GBE1 (F-20): sc-99901

BACKGROUND

GBE1 (glucan (1,4- α -), branching enzyme 1) 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 α -1,4-linked glucosyl units from the outer end of a glycogen chain to an α -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.

REFERENCES

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4. Chan, Y.J., et al. 1999. Glycogen storage disease type IV: a case report. *Zhonghua Yi Xue Za Zhi* 62: 743-747.
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CHROMOSOMAL LOCATION

Genetic locus: GBE1 (human) mapping to 3p12.2; Gbe1 (mouse) mapping to 16 C2.

SOURCE

GBE1 (F-20) is an affinity purified rabbit polyclonal antibody raised against a peptide mapping within an internal region of GBE1 of human origin.

PRODUCT

Each vial contains 100 μ g IgG in 1.0 ml of PBS with < 0.1% sodium azide and 0.1% gelatin.

Blocking peptide available for competition studies, sc-99901 P, (100 μ g peptide in 0.5 ml PBS containing < 0.1% sodium azide and 0.2% BSA).

APPLICATIONS

GBE1 (F-20) is recommended for detection of GBE1 of mouse, rat and human origin by Western Blotting (starting dilution 1:200, dilution range 1:100-1:1000), immunofluorescence (starting dilution 1:50, dilution range 1:50-1:500) and solid phase ELISA (starting dilution 1:30, dilution range 1:30-1:3000).

GBE1 (F-20) is also recommended for detection of GBE1 in additional species, including equine and canine.

Suitable for use as control antibody for GBE1 siRNA (h): sc-78413, GBE1 siRNA (m): sc-145348, GBE1 shRNA Plasmid (h): sc-78413-SH, GBE1 shRNA Plasmid (m): sc-145348-SH, GBE1 shRNA (h) Lentiviral Particles: sc-78413-V and GBE1 shRNA (m) Lentiviral Particles: sc-145348-V.

Molecular Weight of GBE1: 80 kDa.

RECOMMENDED SECONDARY REAGENTS

To ensure optimal results, the following support (secondary) reagents are recommended: 1) Western Blotting: use goat anti-rabbit IgG-HRP: sc-2004 (dilution range: 1:2000-1:100,000) or Cruz Marker™ compatible goat anti-rabbit IgG-HRP: sc-2030 (dilution range: 1:2000-1:5000), Cruz Marker™ Molecular Weight Standards: sc-2035, TBS Blotto A Blocking Reagent: sc-2333 and Western Blotting Luminol Reagent: sc-2048. 2) Immunofluorescence: use goat anti-rabbit IgG-FITC: sc-2012 (dilution range: 1:100-1:400) or goat anti-rabbit IgG-TR: sc-2780 (dilution range: 1:100-1:400) with UltraCruz™ Mounting Medium: sc-24941.

STORAGE

Store at 4° C, **DO NOT FREEZE**. Stable for one year from the date of shipment. Non-hazardous. No MSDS required.

RESEARCH USE

For research use only, not for use in diagnostic procedures.

PROTOCOLS

See our web site at www.scbt.com or our catalog for detailed protocols and support products.