GBE1 polyclonal antibody

Catalog: BS8283 Host: Rabbit Reactivity: Human,Mouse,Rat

BackGround:

GBE1 (glucan (1,4-alpha-), 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 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.

Product:

1mg/ml in PBS with 0.1% Sodium Azide, 50% Glycerol.

Molecular Weight:

~ 80 kDa

Swiss-Prot:

Q04446

Purification&Purity:

The antibody was affinity-purified from rabbit antiserum by affinity-chromatography using epitope-specific immunogen and the purity is > 95% (by SDS-PAGE).

Applications:

WB 1:500 - 1:2000

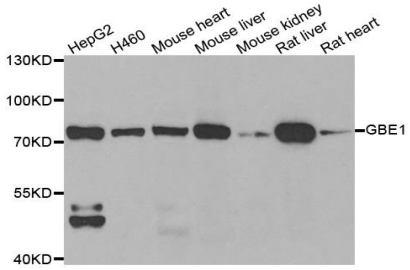
Storage&Stability:

Store at 4℃ short term. Aliquot and store at -20℃ long term. Avoid freeze-thaw cycles.

Specificity:

GBE1 polyclonal antibody detects endogenous levels of GBE1 protein.

DATA:



WesternBlot (WB) analysis of GBE1 polyclonal antibody

Note:

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

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