

bs-13300R**[Primary Antibody]****GBE1 Rabbit pAb**

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— DATASHEET —

Host: Rabbit	Isotype: IgG	Applications: WB (1:500-2000) IHC-P (1:100-500) IHC-F (1:100-500) IF (1:100-500) ICC/IF (1:100-500) ELISA (1:5000-10000) Reactivity: (predicted: Human, Mouse, Rat, Rabbit) Predicted MW.: 80 kDa Subcellular Location: Cytoplasm
Clonality: Polyclonal		
GeneID: 2632	SWISS: Q04446	
Target: GBE1		
Immunogen: KLH conjugated synthetic peptide derived from human GBE1: 101-200/702.		
Purification: affinity purified by Protein A		
Concentration: 1mg/ml		
Storage: 0.01M TBS (pH7.4) with 1% BSA, 0.02% Proclin300 and 50% Glycerol. Shipped at 4°C. Store at -20°C for one year. Avoid repeated freeze/thaw cycles.		
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.		