

bs-13254R**[Primary Antibody]****GAA Rabbit pAb****BioSS**
ANTIBODIES

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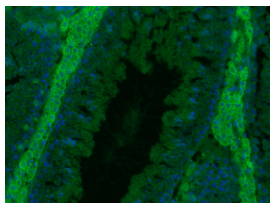
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— DATASHEET —

Host: Rabbit	Isotype: IgG	Applications: IHC-P (1:100-500) IHC-F (1:100-500) IF (1:200-800) Reactivity: Human, Mouse (predicted: Rat, Pig, Sheep, Dog, Horse) Predicted MW.: 83/92/98 kDa Subcellular Location: Cell membrane ,Cytoplasm
Clonality: Polyclonal		
GeneID: 2548	SWISS: P10253	
Target: GAA		
Immunogen: KLH conjugated synthetic peptide derived from human GAA/Glucosidase alpha: 541-640/952.		
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: This gene encodes acid alpha-glucosidase, which is essential for the degradation of glycogen to glucose in lysosomes. Different forms of acid alpha-glucosidase are obtained by proteolytic processing. Defects in this gene are the cause of glycogen storage disease II, also known as Pompe's disease, which is an autosomal recessive disorder with a broad clinical spectrum. Three transcript variants encoding the same protein have been found for this gene. [provided by RefSeq, Jul 2008].		

— VALIDATION IMAGES —

Paraformaldehyde-fixed, paraffin embedded (Mouse testis); Antigen retrieval by boiling in sodium citrate buffer (pH6.0) for 15min; Block endogenous peroxidase by 3% hydrogen peroxide for 20 minutes; Blocking buffer (normal goat serum) at 37°C for 30min; Antibody incubation with (GAA) Polyclonal Antibody, Unconjugated (bs-13254R) at 1:400 overnight at 4°C, followed by a conjugated Goat Anti-Rabbit IgG antibody (bs-0295G-FITC) for 90 minutes, and DAPI for nuclei staining.

— SELECTED CITATIONS —

- **[IF=5.23]** Rauniyar et al. Quantitative Proteomics of Human Fibroblasts with I1061T Mutation in Niemann-Pick C1 (NPC1) Protein Provides Insights into the Disease Pathogenesis. (2015) Mol.Cell.Proteomics. 14:1734-49 WB ;Human. 25873482