bs-13254R

[Primary Antibody]

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GAA Rabbit pAb

DATASHEET -

Host: Rabbit Isotype: IgG

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].

Applications: IHC-P (1:100-500)

IHC-F (1:100-500) **IF** (1:200-800)

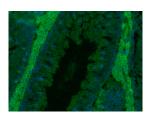
Reactivity: Mouse (predicted: Human,

Rat, Pig, Sheep, Dog, Horse)

Predicted 83/92/98 kDa

Subcellular Location: Cell membrane ,Cytoplasm

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