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GAA Rabbit pAb

货号: **AYP13069**

产品信息

反应	Human,Mouse,Rat
宿主	Rabbit
克隆性	Polyclonal
预测反应	WB: Mus musculus
应用	WB IF/ICC IP
推荐浓度	WB: 1:500 - 1:1000 IF/ICC: 1:50 - 1:200 IP: 1:50 - 1:200
理论分子量	105kDa
实测分子量	105KDa
形式	Liquid
保存条件	Store at -20°C. Avoid freeze / thaw cycles. Buffer: PBS with 0.01% thiomersal,50% glycerol,pH7.3.
偶联物	Unconjugated
阳性对照	DU145,Mouse liver,Mouse ovary,Mouse brain,Rat brain
细胞定位	Lysosome,Lysosome membrane
纯化	Affinity purification

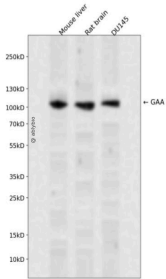
抗原信息

抗原信息	A synthetic peptide corresponding to a sequence within amino acids 350-450 of human GAA (NP_000143.2).
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靶点信息

研究背景	This gene encodes lysosomal alpha-glucosidase, which is essential for the degradation of glycogen to glucose in lysosomes. The encoded preprotein is proteolytically processed to generate multiple intermediate forms and the mature form of the enzyme. 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. Alternative splicing results in multiple transcript variants.
基因ID	2548
基因名	GAA
Swiss	P10253 (https://www.uniprot.org/uniprotkb/P10253/entry)
别名	GAA,LYAG,GAA Rabbit pAb,Acid maltase,Aglucosidase alfa

产品验证



Western blot analysis of GAA expressed in Mouse liver, Rat brain, DU145 using GAA Rabbit pAb at 1:1000. Secondary antibody: HRP Goat Anti-Rabbit IgG (H+L) at 1:5000. Lysates/proteins: 30ug per lane. Blocking buffer: 5% non-fat dry milk in TBST. Detection: ECL Enhanced Kit. Exposure time: 120s.

实验步骤

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