Recombinant GBA Monoclonal Antibody

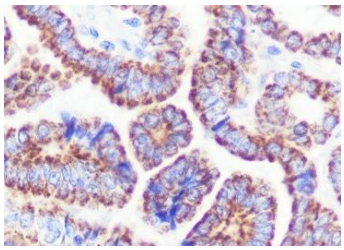
catalog number: AN301962L

Note: *Centrifuge before opening to ensure complete recovery of vial contents.*

Description

Reactivity	Human;Rat;Mouse
Immunogen	Recombinant human GBA fragment
Host	Rabbit
Isotype	IgG, κ
Clone	A678
Purification	Protein A purified
Buffer	PBS, 50% glycerol, 0.05% Proclin 300, 0.05% protein protectant.

Applications	Recommended Dilution
WB	1:1000-1:20000
IHC	1:50



Immunohistochemistry of paraffin-embedded Human thyroid cancer using GBA Monoclonal Antibody at dilution of 1:50.

Preparation & Storage

Storage	Store at -20°C Valid for 12 months. Avoid freeze / thaw cycles.
Shipping	Ice bag

Background

Beta-Glucosylceramidase (β-GC) is a lysosomal enzyme that catalyzes the hydrolysis of glucocerebroside into free ceramide and glucose. Lysosomal breakdown of glucocerebroside is required for cellular metabolism of complex lipids and proper turnover of cellular membrane. In the absence of GBA, the gene that encodes β-GC, autophagic lysosome reformation is altered, suggesting that β-GC activity is critical to maintain functional lysosomes. The cellular function of lysosomes is to degrade and recycle cellular waste to maintain proper cellular energy metabolism. Mutations in human GBA cause deficiency in β-GC, resulting in the accumulation of lysosomal glucocerebroside. Macrophages are particularly sensitive to lysosomal glucocerebroside accumulation due to their role in phagocytosis-mediated breakdown of cellular debris and dying cells. Gaucher disease, a rare autosomal recessive lysosomal storage disorder that is genetically linked to GBA, is marked by engorged “Gaucher cell” macrophages in the spleen, liver, and bone marrow. GBA mutations are the most common genetic risk factor for Parkinson’s disease (PD), a neurodegenerative disease characterized by the loss of dopaminergic neurons in the substantia nigra with formation of α-synuclein-rich Lewy bodies in surviving neurons. GBA mutations may play a direct role in accumulation of α-synuclein by mechanisms that are poorly understood, but may include mislocalization of lysosomal β-GC causing impaired lysosomal degradation of α-synuclein.

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