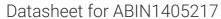
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## anti-GNE antibody (Alexa Fluor 555)



| $\sim$ |     |     |     |
|--------|-----|-----|-----|
|        | N/P | r\/ | i⊢₩ |

| Quantity:    | 100 μL  |
|--------------|---|
| Target:      | GNE   |
| Reactivity:  | Human, Mouse, Rat   |
| Host:        | Rabbit  |
| Clonality:   | Polyclonal  |
| Conjugate:   | This GNE antibody is conjugated to Alexa Fluor 555                              |
| Application: | Western Blotting (WB), Immunofluorescence (Paraffin-embedded Sections) (IF (p)) |
|              |   |

## **Product Details**

| Immunogen:        | KLH conjugated synthetic peptide derived from human GLCNE |
|-------------------|---|
| Isotype:          | IgG   |
| Cross-Reactivity: | Human, Mouse, Rat   |
| Purification:     | Purified by Protein A.                                    |

| Target Details    |  |  |
|-------------------|--|--|
| Target:           | GNE  |  |
| Alternative Name: | GLCNE (GNE Products)   |  |
| Background:       | Synonyms: IBM2, Uae1, Bunctional UDP N acetylglucosamine 2 epimerase/N acetylmannosamine kinase, DMRV, ManAc kinase, N acylmannosamine kinase, NM, RP23- |  |
|                   | 209M8.6, UDP GlcNAc 2 epimerase, UDP GlcNAc 2 epimerase/ManAc kinase, Uridine diphosphate N acetylglucosamine 2 epimerase, GLCNE_HUMAN.                  |  |

Background: The bifunctional enzyme UDP-N-acetylglucosamine 2-epimerase/N-acetylmannosamine kinase (GNE/Mnk), or GLCNE, regulates and initiates biosynthesis of N-acetylneuraminic acid (NeuAc), a precursor of sialic acids. GLCNE is required for normal sialylation in hematopoietic cells. Sialylation is implicated in cell adhesion, signal transduction, tumorigenicity and metastatic behavior of malignant cells. It is upregulated after PKC-dependent phosphorylation and is most abundantly expressed in liver and placenta. It is also expressed, to a lesser extent, in heart, brain, lung, kidney, skeletal muscle and pancreas. Defects in GLCNE are the cause of sialuria, inclusion body myopathy 2 (IBM2) and Nonaka myopathy (NM) or distal myopathy with rimmed vacuoles (DMRV). Sialuria is an autosomal dominant disorder caused by a lack of feedback inhibition of GLCNE by CMP-NeuAc, resulting in overproduction of NeuAc. It is characterized by an accumulation of free sialic acid in the cytoplasm and large quantities of neuraminic acid in the urine. Both IBM2 and NM/DMRV are autosomal recessive neuromuscular disorders characterized by adult onset, distal and proximal muscle weakness (especially in the legs) and a typical muscle pathology including filamentous inclusions and rimmed vacuoles.

Gene ID:

10020

## **Application Details**

| Application Notes: | IF(IHC-P) 1:50-200    |
|--------------------|-----------------------|
| Restrictions:      | For Research Use only |

### Handling

| Format:            | Liquid   |
|--------------------|--|
| Concentration:     | 1 μg/μL  |
| Buffer:            | Aqueous buffered solution containing 0.01M TBS (pH 7.4) with 1 % BSA, 0.03 % Proclin300 and 50 % Glycerol.         |
| Preservative:      | ProClin  |
| Precaution of Use: | This product contains ProClin: a POISONOUS AND HAZARDOUS SUBSTANCE, which should be handled by trained staff only. |
| Storage:           | -20 °C   |
| Storage Comment:   | Store at -20°C. Aliquot into multiple vials to avoid repeated freeze-thaw cycles.                                  |
| Expiry Date:       | 12 months  |