

DESCRIPTION

Species Reactivity	Human
Specificity	Detects human Sulfamidase/SGSH in direct ELISAs.
Source	Monoclonal Mouse IgG _{2A} Clone # 1018387
Purification	Protein A or G purified from hybridoma culture supernatant
Immunogen	<i>Spodoptera frugiperda</i> , Sf 21 (baculovirus)-derived human Sulfamidase/SGSH protein Arg23-Leu502 Accession # P51688
Formulation	Lyophilized from a 0.2 µm filtered solution in PBS with Trehalose. See Certificate of Analysis for details. *Small pack size (-SP) is supplied either lyophilized or as a 0.2 µm filtered solution in PBS.

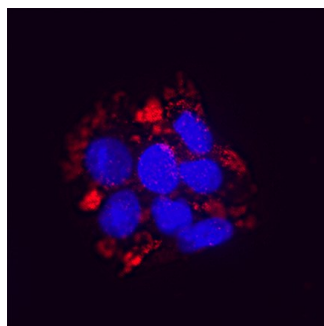
APPLICATIONS

Please Note: Optimal dilutions should be determined by each laboratory for each application. [General Protocols](#) are available in the Technical Information section on our website.

	Recommended Concentration	Sample
Immunocytochemistry	8-25 µg/mL	See Below

DATA

Immunocytochemistry



Sulfamidase/SGSH in HT-29 Human Cell Line. Sulfamidase/SGSH was detected in immersion fixed HT-29 human colon adenocarcinoma cell line (positive stain) and U937 human histiocytic lymphoma cell line (negative stain) using Mouse Anti-Human Sulfamidase/SGSH Monoclonal Antibody (Catalog # MAB8380) at 8 µg/mL for 3 hours at room temperature. Cells were stained using the NorthernLights™ 557-conjugated Anti-Mouse IgG Secondary Antibody (red; Catalog # NL007) and counterstained with DAPI (blue). Specific staining was localized to cytoplasm. View our protocol for [Fluorescent ICC Staining of Cells on Coverslips](#).

PREPARATION AND STORAGE

Reconstitution	Reconstitute at 0.5 mg/mL in sterile PBS.
Shipping	The product is shipped at ambient temperature. Upon receipt, store it immediately at the temperature recommended below. *Small pack size (-SP) is shipped with polar packs. Upon receipt, store it immediately at -20 to -70 °C
Stability & Storage	Use a manual defrost freezer and avoid repeated freeze-thaw cycles. <ul style="list-style-type: none"> 12 months from date of receipt, -20 to -70 °C as supplied. 1 month, 2 to 8 °C under sterile conditions after reconstitution. 6 months, -20 to -70 °C under sterile conditions after reconstitution.

BACKGROUND

Also known as N-sulfoglucosamine sulfohydrolase and heparan N-sulfatase, Sulfamidase/SGSH is an important member of the sulfatase family involved in the degradation of heparan sulfate (HS) (1). Different from the HS specific endosulfatases that remove sulfate from internal GlcNAc residues (2), SGSH removes sulfate group from the non-reducing end glucosamine residues on HS. The SGSH deficiency results in mucopolysaccharidosis type IIIA (MPS IIIA, Sanfilippo A syndrome), an autosomal recessive lysosomal storage disease characterized by neurological dysfunction but relatively mild somatic manifestations (3). Human SGSH shows 88.6% sequence identity with that of mouse sequence.

References:

1. DiezRoux, G. and Ballabio, A. *et al.* (2005) Annu. Rev. Genomics Hum. Genet. **6**:355.
2. Morimoto-Tomita, M. *et al.* (2002) J. Biol. Chem. **277**:49175.
3. Blanch, L. *et al.* (1997) Hum. Mol. Genet. **6**:787.