SunLong Biotech Co.,LTD
Tel: 0086-571- 56623320 Fax:0086-571- 56623318
E-mail:sales@sunlongbiotech.com
www.sunlongbiotech.com

# Rabbit Anti-SGSH antibody

SL11756R

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| **Product Name:** | SGSH |
| **Chinese Name:** | 磺氨基葡糖硫酸胺酶抗体 |
| **Alias:** | Heparan sulfate sulfatase; Heparan sulphate sulphatase; HSS; MPS 3A; MPS3 A; MPS3A; N sulfoglucosamine sulfohydrolase (sulfamidase); N-sulphoglucosamine sulphohydrolase; SFMD; SGSH; SPHM_HUMAN; Sulfoglucosamine sulfamidase; Sulphamidase; Sulphoglucosamine sulphamidase. |
| **Organism Species:** | Rabbit |
| **Clonality:** | Polyclonal |
| **React Species:** | Human,Mouse,Rat,Dog, |
| **Applications:** | WB=1:500-2000ELISA=1:500-1000IHC-P=1:400-800IHC-F=1:400-800ICC=1:100-500IF=1:100-500(Paraffin sections need antigen repair)<br>not yet tested in other applications.<br>optimal dilutions/concentrations should be determined by the end user. |
| **Molecular weight:** | 55kDa |
| **Cellular localization:** | cytoplasmic |
| **Form:** | Lyophilized or Liquid |
| **Concentration:** | 1mg/ml |
| **immunogen:** | KLH conjugated synthetic peptide derived from human Sulphamidase:301-388/502 |
| **Lsotype:** | IgG |
| **Purification:** | affinity purified by Protein A |
| **Storage Buffer:** | 0.01M TBS(pH7.4) with 1% BSA, 0.03% Proclin300 and 50% Glycerol. |
| **Storage:** | Store at -20 °C for one year. Avoid repeated freeze/thaw cycles. The lyophilized antibody is stable at room temperature for at least one month and for greater than a year when kept at -20°C. When reconstituted in sterile pH 7.4 0.01M PBS or diluent of antibody the antibody is stable for at least two weeks at 2-4 °C. |
| **PubMed:** | PubMed |
| **Product Detail:** | Sulfatases are enzymes that hydrolyse a diverse range of sulfate esters. Deficiency of lysosomal sulfatases leads to human diseases characterized by the accumulation of either GAGs (glycosaminoglycans) or sulfolipids. Sulfamidase, also known as HSS, SFMD, MPS3A or SGSH, is a 502 amino acid lysosome that belongs to the sulfatase family. It |

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has been suggested that sulfamidase may be involved in the lysosomal degradation of heparan sulfate. Defects in the gene encoding sulfamidase are the cause of Sanfilippo syndrome A, an autosomal recessive lysosomal storage disease caused by impaired degradation of heparan sulfate. Sanfilippo syndrome A is characterized by severe central nervous system degeneration but relatively mild somatic manifestations.

**Subcellular Location:**
Lysosome.

**Post-translational modifications:**
The conversion to 3-oxoalanine (also known as C-formylglycine, FGly), of a serine or cysteine residue in prokaryotes and of a cysteine residue in eukaryotes, is critical for catalytic activity.

**DISEASE:**
Defects in SGSH are the cause of mucopolysaccharidosis type 3A (MPS3A) [MIM:252900]; also known as Sanfilippo syndrome A. MPS3A is a severe form of mucopolysaccharidosis type 3, an autosomal recessive lysosomal storage disease due to impaired degradation of heparan sulfate. MPS3 is characterized by severe central nervous system degeneration, but only mild somatic disease. Onset of clinical features usually occurs between 2 and 6 years; severe neurologic degeneration occurs in most patients between 6 and 10 years of age, and death occurs typically during the second or third decade of life. MPS3A is characterized by earlier onset, rapid progression of symptoms and shorter survival.

**Similarity:**
Belongs to the sulfatase family.

**SWISS:**
P51688

**Gene ID:**
6448

**Database links:**

Entrez Gene: 6448 Human

Omim: 605270 Human

SwissProt: P51688 Human

Unigene: 31074 Human

**Important Note:**

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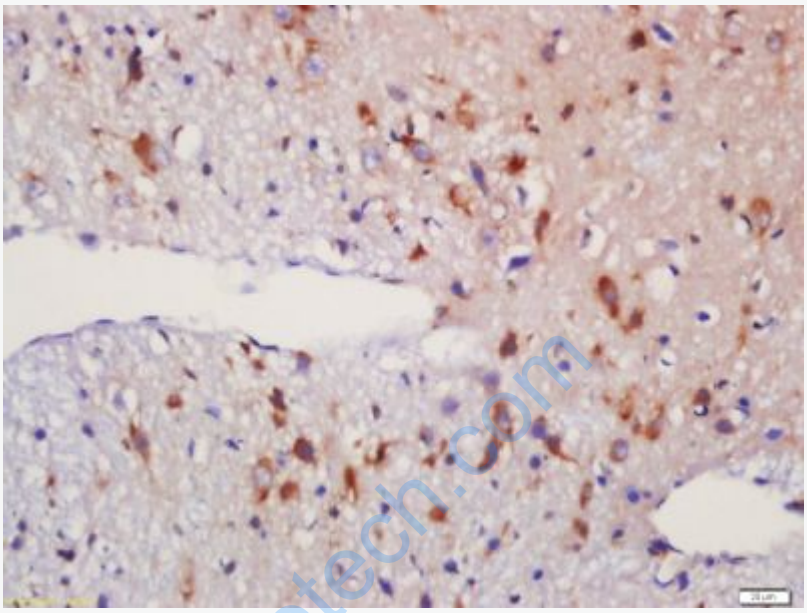
| | This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications. |
| **Picture:** |  <br>Tissue/cell: rat brain tissue; 4% Paraformaldehyde-fixed and paraffin-embedded;<br>Antigen retrieval: citrate buffer ( 0.01M, pH 6.0 ), Boiling bathing for 15min; Block endogenous peroxidase by 3% Hydrogen peroxide for 30min; Blocking buffer (normal goat serum,C-0005) at 37℃ for 20 min;<br>Incubation: Anti-SGSH Polyclonal Antibody, Unconjugated(SL11756R) 1:200, overnight at 4°C, followed by conjugation to the secondary antibody(SP-0023) and DAB(C-0010) staining |

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