10320 Camino Santa Fe, Suite G
San Diego, CA 92121
Tel: 858.875.1900 Fax: 858.875.1999

SGSH Antibody

Purified Rabbit Polyclonal Antibody (Pab)
Catalog # AP50865

Product Information

Application	WB, IHC-P, IHC-F, IF, ICC, E
Primary Accession	P51688
Reactivity	Human, Mouse, Rat, Dog
Host	Rabbit
Clonality	Polyclonal
Calculated MW	56695
Physical State	Liquid
Immunogen	KLH conjugated synthetic peptide derived from human Sulphamidase
Epitope Specificity	301-388/502
Isotype	IgG
Purity	affinity purified by Protein A
Buffer	0.01M TBS (pH7.4) with 1% BSA, 0.02% Proclin300 and 50% Glycerol.
SUBCELLULAR LOCATION	Lysosome.
SIMILARITY	Belongs to the sulfatase family.
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.
Important Note	This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.
Background Descriptions	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 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.

Additional Information

Gene ID	6448
Other Names	N-sulphoglucosamine sulphohydrolase, Sulfoglucosamine sulfamidase, Sulphamidase, SGSH, HSS
Dilution	WB=1:500-2000,IHC-P=1:100-500,IHC-F=1:100-500,ICC=1:100-500,IF=1:100-500,ELISA=1:5000-10000
Format	0.01M TBS(pH7.4) with 1% BSA, 0.09% (W/V) sodium azide and 50% Glyce
Storage	Store at -20 °C for one year. Avoid repeated freeze/thaw cycles. 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.

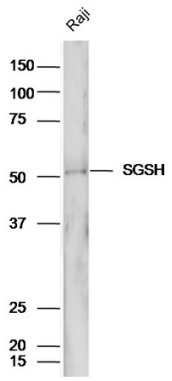
Protein Information

Name	SGSH
Synonyms	HSS
Function	Catalyzes a step in lysosomal heparan sulfate degradation.
Cellular Location	Lysosome.

References

Scott H.S.,et al.Nat. Genet. 11:465-467(1995).
Karageorgos L.E.,et al.Submitted (NOV-1996) to the EMBL/GenBank/DDBJ databases.
Ota T.,et al.Nat. Genet. 36:40-45(2004).
Zhang H.,et al.Nat. Biotechnol. 21:660-666(2003).
Chen R.,et al.J. Proteome Res. 8:651-661(2009).

Images



Raji cell lysates probed with Anti-SGSH/Sulphamidase Polyclonal Antibody, Unconjugated AP50865 at 1:300 in 4˚C. Followed by conjugation to secondary antibody at 1:3000 90min in 37˚C.