

B4GALNT1/GM2 synthase Rabbit pAb

Catalog # AP99623

Product Information

Application	WB, IHC-P, IHC-F, IF
Primary Accession	Q00973
Other Accession	Q00973
Reactivity	Rat, Human, Mouse
Predicted	Rabbit, Dog, Horse, Sheep
Host	Rabbit
Clonality	Polyclonal
Calculated MW	58882
Physical State	Liquid
Immunogen	KLH conjugated synthetic peptide derived from human B4GALNT1/GM2 synthase
Epitope Specificity	451-533/533
Isotype	IgG
Purity	affinity purified by Protein A
Buffer	0.01M TBS (pH7.4) with 1% BSA, 0.02% Proclin300 and 50% Glycerol.
SIMILARITY	Belongs to the glycosyltransferase 2 family.
SUBUNIT	Homodimer; disulfide-linked.
DISEASE	Spastic paraplegia 26, autosomal recessive (SPG26) [MIM:609195]: A form of spastic paraplegia, a neurodegenerative disorder characterized by a slow, gradual, progressive weakness and spasticity of the lower limbs. Rate of progression and the severity of symptoms are quite variable. Initial symptoms may include difficulty with balance, weakness and stiffness in the legs, muscle spasms, and dragging the toes when walking. In some forms of the disorder, bladder symptoms (such as incontinence) may appear, or the weakness and stiffness may spread to other parts of the body. SPG26 is a complicated form characterized by onset in the first 2 decades of life of gait abnormalities due to lower limb spasticity and muscle weakness. Some patients have upper limb involvement. Additional features include intellectual disability, peripheral neuropathy, dysarthria, cerebellar signs, extrapyramidal signs, and cortical atrophy. The disorder is slowly progressive. Note=The disease is caused by mutations affecting the gene represented in this entry.
Important Note	This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.
Background Descriptions	GM2 and GD2 gangliosides are sialic acid-containing glycosphingolipids. GalNAc-T is the enzyme involved in the biosynthesis of G(M2) and G(D2) glycosphingolipids. GalNAc-T catalyzes the transfer of GalNAc into G(M3) and G(D3) by a beta-1,4 linkage, resulting in the synthesis of G(M2) and G(D2), respectively. Three transcript variants encoding different isoforms have been found for this gene. [provided by RefSeq, Feb 2013]

Additional Information

Gene ID 2583

Other Names	GALGT; GALNACT; GalNAc-T; SPG26; 4933429D13Rik; Gal-NAc-T; Galgt1; Ggm-2; Ggm2; B4GN1_HUMAN; B4GALNT1; (N-acetylneuraminyl)-galactosylglucosylceramide; GM2/GD2 synthase; 2.4.1.92; SIAT2; B4GN1_MOUSE; B4GN1_RAT; GA2 synthase; GD2 synthase; GM2 synthase; beta-1, 4-N-acetyl-galactosaminyltransferase 1; UDP-N-acetyl-alpha-D-galactosamine:(N-acetylneuraminyl)-galactosylglucosylceramide N-acetylgalactosaminyltransferase (GalNAc-T); UDP-Gal:betaGlcNAc beta-1, 4-N-acetylgalactosaminyltransferase transferase 1; spastic paraplegia 26; GD2 synthase, GM2 synthase
Dilution	WB=1:500-2000,IHC-P=1:100-500,IHC-F=1:100-500,IF=1:100-500
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	B4GALNT1 (HGNC:4117)
Synonyms	GALGT, SIAT2
Function	Involved in the biosynthesis of gangliosides GM2, GD2, GT2 and GA2 from GM3, GD3, GT3 and GA3, respectively.
Cellular Location	Golgi apparatus membrane {ECO:0000250 UniProtKB:Q10468}; Single-pass type II membrane protein

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