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

CHST6 Rabbit pAb

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Catalog # AP55349

Product Information

Application	WB
Primary Accession	Q9GZX3
Reactivity	Human
Predicted	Dog, Horse, Sheep
Host	Rabbit
Clonality	Polyclonal
Calculated MW	44099
Physical State	Liquid
Immunogen	KLH conjugated synthetic peptide derived from human CHST6
Epitope Specificity	101-200/395
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	Golgi apparatus membrane.
DISEASE	Defects in CHST6 are the cause of macular dystrophy, corneal, 1 (MCDC1) [MIM:217800]. An ocular disease characterized by bilateral, progressive corneal opacification, and reduced corneal sensitivity. Onset occurs in the first decade, usually between ages 5 and 9. Painful attacks with photophobia, foreign body sensations, and recurrent erosions occur in most patients. The disease is due to deposition of an unsulfated keratan sulfate both within the intracellular space (within the keratocytes and endothelial cells) and in the extracellular corneal stroma. Macular corneal dystrophy is divided into the clinically indistinguishable types I, IA, and II based on analysis of the normally sulfated, or antigenic, keratan sulfate levels in serum and immunohistochemical evaluation of the cornea. Patients with types I and IA macular corneal dystrophy have undetectable serum levels of antigenic keratan sulfate, whereas those with type II macular corneal dystrophy have normal or low levels, depending on the population examined. Note=CHST6 homozygous missense mutations have been observed in patients with macular corneal dystrophy type I, while type II patients show a large deletion and replacement in the upstream region of CHST6. The only missense mutation for type II is Cys-50, which is heterozygous with a replacement in the upstream region on the other allele of CHST6.
Important Note	This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.
Background Descriptions	The protein encoded by this gene is an enzyme that catalyzes the transfer of a sulfate group to the GlcNAc residues of keratan. Keratan sulfate helps maintain corneal transparency. Defects in this gene are a cause of macular corneal dystrophy (MCD). [provided by RefSeq, Jan 2010]

Additional Information

Gene ID	4166
Other Names	Carbohydrate sulfotransferase 6, Corneal N-acetylglucosamine-6-O-sulfotransferase, C-GlcNAc6ST, hCGn6ST, 2.8.2.21, Galactose/N-acetylglucosamine/N-acetylglucosamine 6-O-sulfotransferase 4-beta, GST4-beta, N-acetylglucosamine 6-O-sulfotransferase 5, GlcNAc6ST-5, Gn6st-5, CHST6 (HGNC:6938)
Dilution	WB=1:500-2000
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	CHST6 (HGNC:6938)
Function	Sulfotransferase that utilizes 3'-phospho-5'-adenylyl sulfate (PAPS) as sulfonate donor to catalyze the transfer of sulfate to position 6 of non-reducing N-acetylglucosamine (GlcNAc) residues of keratan (PubMed:11278593, PubMed:11352640, PubMed:12218059, PubMed:17690104). Cooperates with B4GALT4 galactosyltransferase and B3GNT7 N-acetylglucosaminyltransferase to construct and elongate the sulfated disaccharide unit [->3Galbeta1->4(6-sulfoGlcNAcbeta)1->] within keratan sulfate polymer. Involved in biosynthesis of keratan sulfate in cornea, with an impact on proteoglycan fibril organization and corneal transparency (PubMed:11278593, PubMed:12218059, PubMed:17690104). Involved in sulfation of endothelial mucins such as GLYCAM1 (PubMed:11352640).
Cellular Location	Golgi apparatus membrane; Single- pass type II membrane protein
Tissue Location	Expressed in cornea. Mainly expressed in brain. Also expressed in spinal cord and trachea

Background

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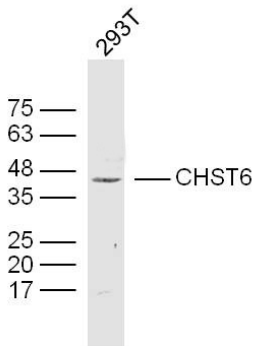
Images



Sample: 293T (human)Cell Lysate at 40 ug
Primary: Anti-CHST6(AP55349) at 1/300 dilution
Secondary: IRDye800CW Goat Anti-Rabbit IgG at 1/20000 dilution
Predicted band size: 44 kD
Observed band size: 44 kD