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GALNS Rabbit pAb

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Catalog # AP55114

Product Information

Application	WB, IHC-P, IHC-F, IF
Primary Accession	P34059
Predicted	Rat, Pig, Dog, Human, Mouse, Horse, Sheep
Host	Rabbit
Clonality	Polyclonal
Calculated MW	58026
Physical State	Liquid
Immunogen	KLH conjugated synthetic peptide derived from human GALNS
Epitope Specificity	1-100/522
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.
DISEASE	Defects in GALNS are the cause of mucopolysaccharidosis type 4A (MPS4A) [MIM:253000]; also known as Morquio A syndrome. MPS4A is a form of mucopolysaccharidosis type 4, an autosomal recessive lysosomal storage disease characterized by intracellular accumulation of keratan sulfate and chondroitin-6-sulfate. Key clinical features include short stature, skeletal dysplasia, dental anomalies, and corneal clouding. Intelligence is normal and there is no direct central nervous system involvement, although the skeletal changes may result in neurologic complications. There is variable severity, but patients with the severe phenotype usually do not survive past the second or third decade of life.
Important Note	This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.
Background Descriptions	Chondroitinase is a 522 amino acid protein that localizes to the lysosome and functions as an exohydrolase that is essential for the degradation of glycosaminoglycans, keratan sulfate and chondroitin 6-sulfate. Using calcium as a cofactor, Chondroitinase, which exists as a disulfide linked oligomer, catalyzes the hydrolysis of the 6-sulfate group on target substrates. Defects in the gene encoding Chondroitinase are the cause of mucopolysaccharidosis type 4A (MPS4A), an autosomal recessive lysosomal storage disease that is characterized by the intracellular accumulation of keratan sulfate and chondroitin-6-sulfate and is associated with dental anomalies, short stature and, in some cases, death in the second or third decade of life.

Additional Information

Gene ID	2588
Other Names	N-acetylgalactosamine-6-sulfatase, 3.1.6.4, Chondroitinsulfatase,

	Chondroitinase, Galactose-6-sulfate sulfatase, GalN6S, N-acetylgalactosamine-6-sulfate sulfatase, GalNAc6S sulfatase, GALNS
Dilution	WB=1:500-2000,IHC-P=1:100-500,IHC-F=1:100-500,ICC/IF=1:100-500,IF=1:100-500,ELISA=1:5000-10000
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	GALNS
Function	Lysosomal enzyme that hydrolyzes sulfate groups from glycosaminoglycans (GAGs), keratan sulfate (KS) and chondroitin-6- sulfate (C6S).
Cellular Location	Lysosome.

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Please note: All products are 'FOR RESEARCH USE ONLY. NOT FOR USE IN DIAGNOSTIC OR THERAPEUTIC PROCEDURES'.