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Lieferung & Zahlungsart

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SANTA CRUZ BIOTECHNOLOGY, INC.

Chondroitinase (h2): 293T Lysate: sc-116653

BACKGROUND

Chondroitinase, also known as GALNS (galactosamine (N-acetyl)-6-sulfate sulfatase), GAS or MPS4A, 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.

REFERENCES

1. Bielicki, J. and Hopwood, J.J. 1991. Human liver N-acetylgalactosamine 6-sulphatase. Purification and characterization. Biochem. J. 279: 515-520.
2. Masue, M., Sukegawa, K., Orii, T. and Hashimoto, T. 1991. N-acetylgalactosamine-6-sulfate sulfatase in human placenta: purification and characteristics. J. Biochem. 110: 965-970.
3. Fukuda, S., Tomatsu, S., Masue, M., Sukegawa, K., Iwata, H., Ogawa, T., Nakashima, Y., Hori, T., Yamagishi, A. and Hanyu, Y. 1992. Mucopolysaccharidosis type IVA. N-acetylgalactosamine-6-sulfate sulfatase exonic point mutations in classical Morquio and mild cases. J. Clin. Invest. 90: 1049-1053.
4. Baker, E., Guo, X.H., Orsborn, A.M., Sutherland, G.R., Callen, D.F., Hopwood, J.J. and Morris, C.P. 1993. The morquio A syndrome (mucopolysaccharidosis IVA) gene maps to 16q24.3. Am. J. Hum. Genet. 52: 96-98.
5. Bielicki, J., Fuller, M., Guo, X.H., Morris, C.P., Hopewood, J.J. and Anson, D.S. 1995. Expression, purification and characterization of recombinant human N-acetylgalactosamine-6-sulphatase. Biochem. J. 311: 333-339.
6. Sukegawa, K., Nakamura, H., Kato, Z., Tomatsu, S., Montano, A.M., Fukao, T., Toietta, G., Tortora, P., Orii, T. and Kondo, N. 2000. Biochemical and structural analysis of missense mutations in N-acetylgalactosamine-6-sulfate sulfatase causing mucopolysaccharidosis IVA phenotypes. Hum. Mol. Genet. 9: 1283-1290.
7. Tomatsu, S., Montano, A.M., Nishioka, T., Gutierrez, M.A., Pena, O.M., Tranda Firescu, G.G., Lopez, P., Yamaguchi, S., Noguchi, A. and Orii, T. 2005. Mutation and polymorphism spectrum of the GALNS gene in mucopolysaccharidosis IVA Morquio A. Hum. Mutat. 26: 500-512.
8. Carraresi, L., Parini, R., Filoni, C., Caciotti, A., Sersale, G., Tomatsu, S., Orlando, C., Zammarchi, E., Guerrini, R., Donati, M.A. and Morrone, A. 2008. GALNS gene expression profiling in Morquio A patients' fibroblasts. Clin. Chim. Acta 397: 72-76.

CHROMOSOMAL LOCATION

Genetic locus: GALNS (human) mapping to 16q24.3.

RESEARCH USE

For research use only, not for use in diagnostic procedures.

PRODUCT

Chondroitinase (h2): 293T Lysate represents a lysate of human Chondroitinase transfected 293T cells and is provided as 100 µg protein in 200 µl SDS-PAGE buffer.

APPLICATIONS

Chondroitinase (h2): 293T Lysate is suitable as a Western Blotting positive control for human reactive Chondroitinase antibodies. Recommended use: 10-20 µl per lane.

Control 293T Lysate: sc-117752 is available as a Western Blotting negative control lysate derived from non-transfected 293T cells.

STORAGE

Store at -20° C. Repeated freezing and thawing should be minimized. Sample vial should be boiled once prior to use. Non-hazardous. No MSDS required.

PROTOCOLS

See our web site at www.scbt.com for detailed protocols and support products.