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ProductInformation

4-Methylumbelliferyl-7-(6-sulfo-2-acetamido-2-deoxy-β-D-glucopyranoside) sodium salt

Product Number **M 0662**
Storage Temperature -0 °C

Product Description

Molecular Formula: $C_{18}H_{20}NNaO_{11}S$
Molecular Weight: 481.4
CAS Number: 142439-99-4
Specific Rotation: -30° (0.5% (w/v) in water)
Extinction Coefficient: E^{mM} = 13.5 (316 nm, water)
Synonyms: MUGS; 4MUGS

MUGS is a sensitive, fluorogenic substrate for β-D-N-acetylglucosaminidase isozyme A activity.[1] The β-hexosaminidases (Hex, EC 3.2.1.52) are lysosomal hydrolases that catalyze the cleavage of terminal β-N-acetylglucosamine or β-N-galactosamine residues on a broad spectrum of glycoconjugates. The major Hex isozymes in humans are: Hex A, a heterodimer composed of one α and one β subunit and Hex B, a homodimer of two β subunits.[2]Both Hex A and Hex B hydrolyze the neutral substrate, 4-Methylumbelliferyl N-acetyl-β-D-glucosaminide, (M 2133). Before the development of MUGS, the assay to differentiate Hex A from Hex B was based on fact that Hex A is more heat labile than Hex B. The Hex A is almost totally inactivated at 50 C, whereas Hex B is stable at this temperature.[3] Hex A is able to hydrolyze both neutral and charged substrates. Hex A is approximately 100 times more active toward MUGS than Hex B.[1] Only Hex A is able to hydrolyze the most important endogenous substrate, the acidic glycolipid GM_2 ganglioside. Mutations in the *HEXA* gene cause Tay-Sachs disease, a GM_2 ganglioside storage disorder.[2,4]

For maximum selectivity of Hex A determination using MUGS substrate, a buffer at pH 4.2 was used.[1] The product of the enzymatic reaction (4-methylumbelliferone) is measured at pH 10.2.[1] 4-Methylumbelliferone has an excitation at 365 nm with emission at 448 nm.[3]

Precautions and Disclaimer

For Laboratory Use Only. Not for drug, household or other uses.

Preparation Instructions

This product is soluble in water (50 mg/ml), yielding a clear, colorless solution. For enzyme assays, it is dissolved at 6 mM (3 mg/ml) in water.

References

1. Bayleran, J., et al., Synthesis of 4-methylumbelliferyl-β-D-N-acetylglucosamine-6-sulfate and its use in classification of GM_2 gangliosidosis genotypes. Clin. Chim. Acta., **143(2)**, 73-89 (1984).
2. Fernandes, M. J. G., et al., Identification of candidate active site residues in lysosomal β-hexosaminidase A. J. Biol. Chem., **272(2)**, 814-820 (1997).
3. Suzuki, K., Enzymatic diagnosis of sphingolipidoses. Meth. Enzymol., **50**, 456-488 (1978).
4. Bayleran, J., et al., Tay-Sachs disease with hexosaminidase A: characterization of the defective enzyme in two patients. Am. J. Hum. Genet. **41(4)**, 532-548 (1987).

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