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ProductInformation

4-Methylumbelliferyl a-D-galactopyranoside

Product Number **M7633** Storage Temperature -0 °C

Product Description

Molecular Formula: $C_{16}H_{18}O_8$ Molecular Weight: 338.3 CAS Number: 38597-12-5 Melting Point: 221-222 °C¹ Specific Rotation: +237° (0.3% (w/v) in water)¹ Extinction Coefficient: $E^{mM} = 14.23$ (318 nm, methanol)

This compound is a substrate for α -galactosidase. It has been used in studies of Fabry's disease in which the enzyme α -galactosidase A is deficient.²⁻⁷ The enzyme is assayed in sodium citrate, pH 4.5, and terminated with glycine buffer, pH 10.8. The released 4-methylumbelliferone is measured fluorometrically with excitation at 365 nm and emission at 448 nm.² 4-Methylumbelliferyl α -D-galactopyranoside continues to be used in the study of α -galactosidase mutants.^{8,9}

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 to faint yellow solution with the application of heat..

References

- 1. Chem. Listy. **52** 1629 (1958).
- Suzuki, K., Enzymatic diagnosis of sphingolipidoses. Meth. Enzymol., 50, 456-488 (1978).
- Beutler, E. and Kuhl, W., Biochemical and electrophoretic studies of α-galactosidase in normal man, in patients with Fabry's disease, and in Equidae. Amer. J. Hum. Genet., **24(3)**, 237-249 (1972).
- Schram, A. W., et al., Enzymological properties and immunological characterization of α-galactosidase isoenzymes from normal and Fabry human liver. Biochim. Biophys. Acta. 482(1), 125-137 (1977).
- Desnick, R. J., et al., Fabry's disease: enzymatic diagnosis of hemizygotes and heterozygotes.
 α-galactosidase activities in plasma, serum, urine, and leukocytes. J. Lab. Clin. Med., 81(2), 157-171 (1973).
- Rietra, P. J., et al., Properties of the residual α-galactosidase activity in the tissues of a Fabry hemizygote. Clin. Chim. Acta. 62(3), 401-413 (1975).
- Bishop, D. F. and Desnick, R. J., Affinity purification of α-galactosidase A from human spleen, placenta, and plasma with elimination of pyrogen contamination. Properties of the purified splenic enzyme compared to other forms. J. Biol. Chem., **256(3)**, 1307-1316 (1981).
- Lemansky, P., et al., Synthesis and processing of α-galactosidase A in human fibroblasts. Evidence for different mutations in Fabry disease. J. Biol. Chem., 262(5), 2062-2065 (1987).
- Kase R., et al., Characterization of two αgalactosidase mutants (Q279E and R301Q) found in an atypical variant of Fabry disease. Biochim. Biophys. Acta., **1501(2-3)**, 227-235 (2000).

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