

CHITOTRIOSIDASE ENZYME : STUDY OF BIOCHEMICAL PARAMETERS IN NORMAL INDIVIDUALS AND PATIENTS WITH GM1-GANGLIOSIDOSIS

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Chitotriosidase (CT) is a family member of glycosylhydrolases with chitin degrading activity. CT activity is usually highly increased in the plasma of patients with Gaucher Disease (GD). A less marked increase in CT activity is also found in GM1-gangliosidosis (GM1). The aim of the present study was to investigate the kinetics of chitotriosidase (CT) in normal individuals and patients with GM₁ Gangliosidosis (GM1). CT activity in GM1 patients was around 14-fold greater than in normal individuals. In patients with GM1, optimum pH for CT was 5.63 ± 0.32 , while in normal individuals the optimum pH was 5.17 ± 0.26 . The mean Km of CT from GM1 patients was $35 \pm 9 \mu\text{M}$ and Vmax was $1222 \pm 615 \text{ nmol/h/mL}$, as compared with Km of $4.7 \pm 1.7 \mu\text{M}$ and Vmax of $79 \pm 61 \text{ nmol/h/mL}$ of normal individuals. We found that the enzyme of normal individuals was more stable to heat, as compared to GM1 patients. The variation of the parameters analyzed may be useful to distinguish normal individuals from patients with GM1 in high-risk population, but it will not replace the need for specific enzyme testing to achieve a final diagnosis of this disease.

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