A-9

Clinica Chimica Acta, 104 (1980) 301–308

CCA 1389

FAMILIAL TEMPERATURE SENSITIVE ALPHA 1 PROTEASE INHIBITOR (M1ANAHEIM)

JOSEPH CHARLES TAYLOR, MONA COLIN, TSUTOMU INAMIZU * and CHARLES MITTMAN

Respiratory Disease Department, City of Hope National Medical Center, 1500 East Duarte Rd., Duarte, CA 91010 (U.S.A.)

(Received November 5th, 1979)

Summary

An α_1 protease inhibitor which is sensitive to increased temperatures and which migrates in the same position as the normal M1 phenotype, has been identified. The sensitivity of the trypsin inhibitory capacity is an inherited characteristic, which is enhanced by acid conditions, as well as conditions during isoelectric focusing. Loss of the serum trypsin inhibitory capacity is not parallel to that of the serum elastase capacity, suggesting that the inhibitory sites for these proteases are oriented differently within the α_1 protease inhibitor molecule.

Introduction

Alpha 1 protease inhibitor (alpha-1-antitrypsin) is one of at least 6 or 8 protease inhibitors identified in human blood [1]. It is the major protease inhibitor (Pi) in serum and is one of the most polymorphic of human proteins. The amount and type of the protease inhibitor found in any individual appears to be controlled at a single autosomal gene locus. Inheritance seems to be codominant; at least 26 different alleles have been described to date. The allele products have been designated by symbols according to the electrophoretic migration velocity of the protein on acid starch gels. Thus, the mobility of the most prevalent type, M1, is moderate, that of F is fast and that of S is slow. The allele associated with alpha 1 protease inhibitor (α_1PI) deficiency, Pi^Z has even slower mobility than the S protein.

In addition to differences in electrophoretic mobility, differences in other

* Visiting Physician from Department of Internal Medicine, Hiroshima University School of Medicine. Correspondence should be sent to J.C. Taylor, Ph.D.