The objective of this study is to examine taurine supplementation (30 mg/Kg/d) effects on serum and fecal BA pattern, and on fat excretion, in CF patients. 23 children, aged 17 months to 19 years, were classified into two groups. GI patients (12) had severe steatorrhea (23.4±4.1 g.24h), and GII were 11 malnourished children with mild fecal fat excretion (4.2±0.7 g.24h). Before taurine and after 4 months of supplementation, patients were monitored for blood and fecal BA (enzimatic-fluorometric method) profiled by thin layer chromatography, nutritional status (assessed by conventional test), and fecal fat excretion (van der Kamer method). After taurine, fasting total serum BA increased from 26.4±7.7 μ mol/l to 48.0±9.9 μ mol/l in GI patients, while in GII children increased from 26.8±6.6 μ mol/l to 38.2±12.6 μ mol/l. Besides glycine/taurine conjugation was altered in all patients. Before taurine, all patients had fecal BA malabsorption (average 688 mg.24h). After supplementation, only GI children showed a remarkable ($p<0.001$) reduction in fecal BA malabsorption, and six of them showed also a significant ($p<0.001$) decrease of fat excretion. In contrast, in all GII patients fat malabsorption was important ($P<0.001$) after taurine. For both GI and GII, after taurine, monitoring of growth revealed a marginal increase of weight in 52% of them, correlated with a steatorrhea reduction in 16% of the children. Results suggest that taurine supplementation can be a useful adjunct form of therapy in CF patients with fat malabsorption. However, the serum BA increase might cause liver damage.

DECREASED VIP RECEPTOR BINDING IN MONONUCLEAR CELLS WITH A GENETIC MEMBRANE FLUIDIZATION.

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Department of Paediatrics, University of Hamburg, FRG. Vasoactive intestinal peptide (VIP) is recognized as a neurotransmitter and immunomodulator. Mononuclear cells possess functional VIP receptors. In HT29 cells specific VIP binding could be modulated by plasma membrane fluidity alterations (El Battari et al., Biochimie 67:1217,1985). Studying membrane alterations in the genetic disease, Myotonic Dystrophy (MD), we found an increased membrane fluidity in MD mononuclear cells as indicated by decreased 1,6-diphenyl-1,3,5-hexatriene fluorescence anisotropy (0.163 ± 0.017 , $x\pm SD$, $n=13$) vs controls (0.181 ± 0.015 , $n=13$, $p<0.01$, U-test). We therefore studied VIP binding to MD cells according to Guerrero et al., Mol Cell Endocrinol 21:151, 1981. Specific binding of VIP to MD cells was reduced ($2.9\pm0.9\%/10^6$ cells, $n=8$; vs controls: $5.2\pm1.6\%/10^6$ cells, $n=9$, $p<0.005$). Binding sites per cell were normal. Scatchard analysis indicated a single class of high affinity binding sites over the range of 10^{-12} to 5×10^{-9} M VIP. Affinity was reduced in MD cells ($K_d = 0.26\pm0.05$ nM vs controls $K_d=0.19\pm0.02$ nM, $p<0.005$). Our data demonstrate that VIP binding is decreased in membranes fluidized by a genetic defect. This could be due to a displacement of membrane embedded VIP receptors supporting the hypothesis of interdependence between receptor function and membrane fluidity.

REGULATION OF SUCRASE-ISOMALTASE MESSENGER RNA CONCENTRATION IN RAT SMALL INTESTINE BY SUCROSE FEEDING.

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Sucrose feeding is known to induce increase of sucrase (S) and isomaltase (I) activities. In order to document the molecular process involved, we studied in rat, the regulation of the transcription of the corresponding mRNA in response to sucrose feeding. In this purpose, a cloned rat S-I probe has been prepared. By Northern hybridization in high stringent conditions, the cDNA sequence hybridizes to a single 6kb nucleotides species specific to the rat intestine.

After 3 days of pre-feeding a carbohydrate free diet (cellulose diet, CD), rats were fed either with CD, either with a diet containing high amounts of carbohydrates (sucrose diet, SD). Animals were then killed at 0, 1h30, 3h, 6h and 15h after refeeding, and a jejunal segment removed. In the mucosal homogenates, a significant increase in S and I activities ($p<0.05$) occurred only 3h after initiation of SD. Northern hybridization of equal amount of total jejunal RNA to rat S-I cDNA probe indicated that after 1h30 of refeeding, SD rats had twice as much S-I mRNA as those on cellulose diet. This difference in S-I mRNA levels was maintained or increased at the other experimental times. In contrast, hybridization with β -actin cDNA failed to show difference in the β -actin mRNA.

The rapid production of SI mRNA provides convincing evidence that the carbohydrate stimulated rise of sucrase activity involved an increase of S-I gene expression at the transcriptional level, associated or not with a stabilization of the S-I messenger.