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The major symptoms in cystic fibrosis (CF) are abnormal epithelial chloride conductance and increased mucus production. These processes are regulated by arachidonic acid (AA) and its products in the eicosanoid system (prostaglandins, leukotrienes, hydroxyeicosatetraenoic acids). An increased release of AA has been demonstrated in lymphocytes and fibroblasts from CF patients due to a defect response to dexamethasone stimulation. This defect might be explained by an abnormal or an absence of a lipocortin, secreted from cells and normally inhibiting phospholipase A₂, the rate limiting enzyme in eicosanoid synthesis. Increased amounts of leukotrienes from white blood cells and increased urinary excretion of most products in the prostanoic system in non-infected CF patients support this hypothesis. The absence of inhibition of AA release ultimately gives an essential fatty acid deficiency EFAD, which in animals have been shown to give increased epithelial sodium transport, fat malabsorption, liver steatosis, increased caloric need and increased bacterial colonization of airways - all wellknown clinical symptoms in CF and some of them shown to be influenced by administration of Intra-lipid[®]. Our hypothesis is that the major symptoms in CF are due to an increased AA release due to a defect in/or absence of lipocortin. The subsequent EFAD gives rise to most of the other clinical symptoms and the progress of the disease. The degree of lipocortin deficiency might determine the rate of development of the disease and the type of presenting symptoms. The wellknown influence of the nutritional status on the progress of the disease will be explained by its relation and capacity to balance the EFAD.

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Dept of Pediatrics, Karolinska Institute, Huddinge Univ Hospital, Stockholm, Dept of Medicine, Univ of Lund, Lund, Sweden. Patients with cystic fibrosis have EFAD which might contribute to their symptoms, especially their poor fat absorption, since rats with EFAD have been shown to have a decreased absorption of fat. To further study the pathophysiology of this defect ³H-arachidonic (20:4) and ¹⁴C-linoleic (18:2) acids were fed intragastrically to EFAD male rats and to age-matched controls. The incorporation into different lipid classes of the small intestine, liver and serum was studied 1-4 hrs after feeding. In both the control and the EFAD rats substantial amounts of ³H-20:4 were retained in the small intestinal and hepatic phospholipids rather than being secreted as lipoprotein triglycerides over the time period studied, the ³H/¹⁴C ratio of the intestinal and hepatic lipids being significantly higher than in controls, (p<0.001). The ³H/¹⁴C ratio of total lipids in intestine and liver was significantly lower in the EFAD than in the controls, due to a marked increase in the retention of ¹⁴C-18:2 in these organs. Also the distribution of the ³H and ¹⁴C between different lipid classes varied markedly between the groups, the proportion of both isotopes in the phospholipids being increased, due to a marked increase in the incorporation of ¹⁴C and ³H into phosphatidylethanolamine (PE) in both liver and intestine (p<0.001) and to some extent of the ¹⁴C into phosphatidylcholine (PC) (p < 0.001). Thus EFAD rats selectively retain absorbed 18:2 and 20:4 in PE and PC of small intestine and liver. The mechanism has to be further studied in view of the importance for intestinal transport and fat malabsorption.

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