



Prevention of Essential Fatty Acids Deficiency as a Part of Nutrition Therapy in Patients with CF

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Survival from cystic fibrosis (CF) has substantially improved over the past four decades. Many factors have contributed to this, including co-ordinated center-based care, earlier neonatal diagnosis, improved airway clearance techniques and application of broad spectrum antibiotics. One of the most important contributing factors leading to better survival and improved quality of life is the advance in nutritional management of CF.

Nutrition starts to be a concern in cystic fibrosis very soon after birth. Newborns with CF are usually smaller than infants of the same age and gender and approximately 85-90% of infants with CF are pancreatic insufficient. Most infants with CF display evidence of steatorrhoea by one year of age. The target of nutritional therapy for patients with CF is to achieve normal growth and physical development, comparable with the healthy population. Although this aim is clear and simple, for its achievement an enormous amount of effort is necessary. For parents of children with CF as well as for patients with CF themselves, it can be like trying to do their best to move up an escalator which is running down.

Patients with CF are at risk of essential fatty acid (EFA) deficiency. Essential fatty acids can not be synthesized in the body so must be obtained from food. A healthy person requires 1-2% of their daily energy intake to be supplied by EFAs. Essential fatty acids are important constituents of cell membranes, influencing membrane permeability and transport functions. They are also important for enzymatic activity and gene expression.

Abnormal levels of EFAs in plasma phospholipids of patients with CF have been reported for more than forty years. In addition, it has been shown that EFA deficiency is present in patients with CF in the absence of protein and energy malnutrition. Reduced availability of EFA may lead to:

- Alteration of mucosal membrane composition
- Impairment of fat transport via across mucosa
- Contribution to malabsorption by interfering with intra-enterocyte lipid transport
- Profound changes in membrane fluidity and cell signalling mechanisms

Although the pathogenesis of EFA deficiency still remains unknown, it is clear that some, clinical symptoms in CF are closely connected with EFA deficiency. These include:

- Desquamating skin lesions
- Increased susceptibility to infections
- Impaired growth
- Hepatic steatosis (*fatty liver*)

For improvement in the health status of patients with CF it is very important to avoid EFA deficiency. The simplest way is to *regularly consume oily fish and* to add one or two



spoons of corn (maize) or soya oil to every day food like rice, pasta, soup or potatoes. In cases of mild deficiency an appropriate EFA supplement may be recommended by your dietitian or physician. If a patient with CF is suffering of severe EFA deficiency an intravenous infusion of Intralipid may be recommended by the physician.