USANA Technical Bulletin

Cystic Fibrosis

Description

- Cystic Fibrosis is an inherited disease of the exocrine glands. It affects the pancreas, respiratory system, intestines, liver and sweat glands. Patients experience chronic respiratory infections, pancreatic insufficiency, and increased electrolytes in sweat. The pancreatic insufficiency leads to nutrient deficiencies, which can further impair the body's ability to combat the respiratory infections.[1]

Causes

- Cystic fibrosis is a genetic disease. It is inherited as an autosomal recessive trait.[2]

Stages

- Cystic fibrosis is a progressive disease and ultimately may require heart and lung transplants.

At Risk

- Children of parents who both carry the recessive gene have a 25% chance of inheriting this disease.

Prevention and Management

- Currently there is no way to cure cystic fibrosis. Medical interventions are intended to help the patient lead as normal a life as possible.
- Obstruction of the pancreatic ducts results in a deficiency of trypsin, amylase and lipase which prevents the conversion and absorption of fat and protein. This interferes with the absorption of fat soluble vitamins, A, D, E and K. Diets should include supplements of these vitamins.[2]
- One study suggests that cystic fibrosis patients have inadequate antioxidant defenses to cope with the increased oxidative stress that these patients regularly experience.[3]
- Efficient antioxidant supplementation may decrease lung inflammation in cystic fibrosis.[4]
- Supplementation may be necessary to maintain normal levels of beta carotene.[5] The observed increased levels of lipid peroxidation may be due to low levels of beta-carotene. These low levels may be increased with supplementation.[6]

- Patients with malabsorption syndromes may develop vitamin E deficiencies and tailored treatment may include vitamin E supplements.[7] Patients supplemented with at least 100 mg of vitamin E had normal concentrations of vitamin E in their erythrocytes. Patients that were unsupplemented or who received amounts of vitamin E less than 100 mg had levels below normal.[8]

Sources of Additional Information

- http://www.cff.org/
- http://ourworld.compuserve.com/homepages/FAntognini/iacfa.htm

Abstract

Lepage G, Champagne J, Ronco N, Lamarre A, Osberg I, Sokol RJ, Roy CC. Supplementation with carotenoids corrects increased lipid peroxidation in children with cystic fibrosis. Am J Clin Nutr 1996 Jul;64(1):87-93. Evidence of lipid peroxidation previously documented in cystic fibrosis (CF) implies an imbalance between free radical generation and antioxidant defense mechanisms. The aim of the present study was to examine the relation between plasma concentrations of malondialdehyde, a marker of lipid peroxidation, and the exogenous antioxidant line of defense. Malondialdehyde concentrations (90.2 +/- 4.7 nmol/L) in 25 children with CF aged 9.6 +/- 0.8 y were higher (P < 0.001) than concentrations (69.1 +/- 2.6 nmol/L) in 17 children used as control subjects and were not correlated with any marker of disease severity. In contrast with their all-rac-alpha-tocopherol status, which was normal as a result of routine supplementation with a 200-mg dose of all-rac-alpha-tocopheryl acetate/d, beta-carotene was very low. A 2-mo open trial in which 12 children with CF aged 11.5 +/- 0.8 y were given 4.42 mg (8.23 mumol) beta-carotene three times per day led to normalization of the malondialdehyde concentration in all but 1 patient, in conjunction with an increase of plasma beta carotene from 0.08 +/- 0.03 to 3.99 +/- 0.92 mumol/L. Their plasma concentrations were inversely correlated (r = -0.54, P = 0.006) [corrected] with malondialdehyde when the values measured pre- and posttreatment were pooled. We conclude that beta-carotene deficiency contributes to lipid peroxidation in CF and that supplementation may eventually prove to be a useful adjunct for the management of the disease.

References

[1] Taber's Cyclopedic Medical Dictionary. 16th ed. Philadelphia:F.A. Davis Company; 1985. p 446.
[2] Diseases. Springhouse (PA):Springhouse Corporation; 1993; p 656-7.
[3] Brown RK et al. Pulmonary dysfunction in cystic fibrosis is associated with oxidative stress. EurRespir J 1996 Feb;9(2):334-9.
[4] Winklhofer-Roob BM et al. Neutrophil elastase/alpha 1-proteinase inhibitor complex levels decrease in plasma of cystic fibrosis patients during long-term oral beta-carotene supplementation. Pediartr Res 1996 Jul;40(1):130-4.
[5] Winklhofer-Roob BM, Response to oral beta-carotene supplementation in patients with cystic fibrosis: a 16-month follow-up study. *Acta Paediatr* Oct;84(10)1132-6.
[6] Lepage G, et al., Suplementation with carotenoids corrects increased lipid peroxidation in children with cystic firosis. *Am J Clin Nutr* 1996 Jul;64(1):87-93.
[7] Tanyel MC; Mancano LD, Neurologic findings in vitamin E dificiency. *Am Fam Physician* 1997 Jan;55(1):197-201.
[8] Perters SA; Kelly FJ, Vitamin E supplementation in cystic fibrosis. *J Pediatr Gastroenterol Nutr 1996 May;22(4):341-5.*