

Serum and urinary carnitine in children with cystic fibrosis

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ABSTRACT

Purpose: Cystic fibrosis (CF) is inherited, congenital disease of multi-organ expression. Carnitine play a role as a lipid acid transporter to mitochondrium for beta-oxydation. Acylation of carnitine is inevitable for detoxication processes in cells. Low lean body mass In CF patients can lead to decreased levels. The aim of the study was the evaluation of free carnitine, acylcarnitine and acylcarnitine/free carnitine ratio in serum and urine of children with cystic fibrosis.

Material and methods: The study was conducted in a group of 15 CF children (4 F, 11 M), aged 12.6 ±5.4 years. The serum for a control group was collected from 32 healthy children. Urine samples for control group was collected from 62 health children. Free carnitine and total carnitine was assessed using spectrophotometric method in which acyl group is transferred from acetyl-CoA to carnitine by carnitine acetyltransferase (CAT). Acyl carnitine concentration and acylcarnitine/free carnitine ratio was counted using Schmidt-

Sommerfeld and Seccombe equation.

Results: In 12 CF patients (80%) free carnitine and total carnitine was below lower limit of normal ($p < 0.001$). In 9 patients (60%) free carnitine level was $\leq 20 \mu\text{mol/L}$, which can be clinically diagnosed. Acylcarnitine levels were also statistically lower in CF group ($p < 0.01$). Acylcarnitine/free carnitine ratio did not differ between the groups ($p = 0.05$). Urine excretion of free carnitine, total carnitine and acylcarnitine was lower in CF group ($p < 0.001$).

Conclusions: In CF pediatric patients statistically significant lower levels of free carnitine, total carnitine and acylcarnitine were observed in comparison to controls. Low urine excretion of free carnitine, total carnitine and acylcarnitine was lower in CF group. No correlation between serum and urine levels of free carnitine, total carnitine and acylcarnitine were observed.

Key words: cystic fibrosis, free carnitine, total carnitine, acyl carnitine
