

Abstract

Eur J Pediatr. 2005 Mar;164(3):131-4.

Serum carnitine levels in patients with homozygous beta thalassemia: a possible new role for carnitine?

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BACKGROUND: Carnitine (beta-hydroxy-gamma-trimethylaminobutyric acid) facilitates the transfer of activated long-chain fatty acids from the cytoplasm to the mitochondria, the site of their beta-oxidation. Carnitine deficiency results in a reduced usage of fatty acids in energy production and therefore the appearance of clinical symptoms such as myalgia and muscle weakness.

METHODS: In the present study, serum carnitine levels were measured in 45 children and 20 adults with homozygous beta thalassemia.

RESULTS: A decrease in serum carnitine levels (total, free and acyl) was found, without any evidence of disorder in the process of mitochondrial beta-oxidation. The possible cause of this finding could be related to a reduced hepatic carnitine biosynthesis.

CONCLUSION: In patients with homozygous beta thalassemia, the reduction of serum carnitine levels might play an important role in the appearance of muscular dysfunction. It is possible that L : -carnitine administration in these patients might improve or even resolve the aforementioned symptom.

PMID: 15717177

