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Case Report

L-Carnitine as a treatment of lethargy in children with chronic neurologic handicaps

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We present five cases of children with severe neurologic handicaps whose management was complicated by excessive lethargy. Treatment with L-carnitine in a dosage range of 35–50 mg/kg/day resulted in a marked improvement in alertness and arousability. In four cases, when L-carnitine was discontinued for a month, they all promptly became lethargic. When L-carnitine was re-started, the lethargy resolved and the improvement has been maintained for up to 14 months. In three children who were tested, serum carnitine levels (total and free) were normal before starting L-carnitine treatment.

Key words: L-Carnitine; Lethargy

1. INTRODUCTION

Carnitine is essential in mitochondrial energy metabolism. It has two principal functions: (i) to transport long-chain fatty acids into the mitochondrion; (ii) to help regulate the intramitochondrial ratio of acetyl-coenzyme A (CoA) to free CoA (reviewed in [1]). Carnitine deficiency conditions may be primary, such as those associated with inborn errors of metabolism, or secondary, such as those associated with inadequate intake or those that are induced by medications. Clinical symptoms of carnitine deficiency may include: myopathy, cardiomyopathy and Reye syndrome-like encephalopathy. In a child with Rett syndrome, with normal total and free serum carnitine levels, L-carnitine treatment resulted in improved neurologic functioning, especially in improvement in her level of alertness [2]. We decided to treat four children who were developmentally disabled and who were excessively lethargic, with L-carnitine to see if their level of alertness could likewise be improved.

2. MATERIALS AND METHODS

2.1. Case 1

This is a 16-year-old female. At the age of 4 months, after receiving a DPT and oral polio immunization, she developed seizures and respiratory insufficiency. Her spinal fluid analysis revealed pleocytosis (30 lymphocytes per mm^3) with normal chemistries and negative cultures. She was treated with ampicillin, isoniazid and streptomycin. EEG tracings revealed hypsarrythymia. She eventually developed microcephaly, spastic quadriparesis and an ongoing seizure disorder which was controlled with valproic acid and carbamazepime. Psychologic evaluation revealed a Bayley Scale of Infant Development IQ of below 10 (profound mental retardation) and Vineland Adaptive Behavior Scales of between 0 and 1 months in communication, daily living skills and socialization. The medications that she was taking were 2,100 mg per day valproic acid, 450 mg per day carbamazepime, and 30 mg per day clorazepate di-potassium for spasticity. Her routine blood chemistries, thyroid function tests, liver function tests,

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