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L-Carnitine

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Editorial

Introduction to the Special Issue on Carnitine

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Carnitine is a relatively simple, ubiquitous molecule that can be considered essential for life because of its critical role in energy metabolism. The essential role of carnitine in metabolism is also reflected in the mechanisms that have evolved to maintain stable carnitine levels. Thus, carnitine is absorbed from the diet, synthesized from dietary precursors, and reabsorbed from the kidney, and compensatory mechanisms exist to adjust the relative contributions of each of these processes to maintain carnitine homeostasis.

The role of carnitine in health and disease has become increasingly apparent, and an extensive biochemical and clinical literature has developed. These studies suggest that an understanding of carnitine is relevant for many areas of clinical medicine, including nutrition, gastroenterology, endocrinology, cardiology, and nephrology. The purpose of this special issue of the *Journal of Child Neurology* is to highlight the relevance of carnitine for child neurology. The peer-reviewed articles included in this special issue discuss aspects of carnitine metabolism and deficiency that child neurologists may encounter in their clinical practice.

Carter, Abney, and Lapp provide the biochemical background necessary to understand the role of carnitine in clinical neurology. They point out that the pathways for carnitine biosynthesis are incompletely developed at birth, so premature and newborn infants are more dependent on dietary sources of carnitine. Borum develops this theme further in her article and notes that neonates on parenteral nutrition are particularly stressed because they cannot make carnitine and are receiving no carnitine in the diet. Carnitine supplementation in these infants seems to promote metabolism and improve growth. Coulter's review of carnitine deficiency in epilepsy also notes that carnitine deficiency is common in preterm infants with seizures, but it is not yet clear that carnitine supplementation will prevent seizures.

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Pons and DeVivo discuss the clinical disorders in which child neurologists may encounter carnitine deficiency. Although many of these are rare, prompt recognition is essential because carnitine treatment can be very effective. They include acquired medical disorders and iatrogenic factors that may induce carnitine deficiency. Perhaps the most common clinical situation in which child neurologists may encounter carnitine deficiency is the treatment of epilepsy. Coulter's article discusses this situation in detail, developing the theme that sufficient data exist to identify risk factors that predict which patients with epilepsy are most likely to have carnitine deficiency. The status of carnitine treatment in epilepsy is also reviewed. Astute readers may note some discrepancy in treatment recommendations between these articles, which reflects alternative clinical responses to incomplete data regarding the effectiveness of carnitine treatment. Clearly, more data are needed to resolve these issues, but these articles provide the currently available data from which readers may draw their own conclusions.

Several other clinical situations arise in which child neurologists may see patients with carnitine deficiency. Child neurologists are increasingly involved in the care of patients with human immunodeficiency virus infection and acquired immune deficiency syndrome (AIDS). Mintz reviews the evidence for carnitine deficiency in these patients. Studies of carnitine treatment in these patients are difficult to do because of the complicated nature of the illness, but evidence is accumulating to support the possible effectiveness of carnitine in AIDS. Winter's review of carnitine in pediatric cardiomyopathy is included in this issue because child neurologists may be asked to evaluate these patients in consultation, and pediatricians' awareness of the role of carnitine in these patients is limited.

Finally, child neurologists should consider several other situations in which carnitine deficiency may occur but for which there are currently few or no data. For example, the data in Coulter's article suggest risk factors to identify carnitine deficiency in patients receiving valproic acid. Because valproic acid is increasingly used in psychiatry to treat children with a variety of disorders including aggression and disorders of mood and affect, the effect of valproic acid on carnitine metabolism may also become apparent in these patients. Valproic acid is

also being used to prevent migraine and could have effects on carnitine metabolism in these patients as well.

The articles included in this special issue should provide the information needed to assist child neurologists in their evaluation and management of these and other patients. The authors hope that these articles will stimulate further research to clarify the role of carnitine deficiency and treatment in pediatrics and neurology.

Note from the Editor-in-Chief

The policy of the *Journal of Child Neurology* is that all editorial materials (including invited articles for supplemental issues) are peer reviewed. For this special supplementary issue on carnitine, the following individuals (in addition to members of the Editorial Board) have graciously assisted in peer review of articles: A. Chadwick Cox, Jeanie B. McMillin, Van S. Miller, and Hugo W. Moser. I also acknowledge the sponsorship of this special issue by Sigma-Tau Pharmaceuticals Inc.