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COMMENTARY

Consideration of betaine and one-carbon sources of N^5-methyltetrahydrofolate for use in homocystinuria and neural tube defects[1,2]

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A major focus in attempts to ameliorate homocystinuria and neural tube defects is supplementation of the diet with B vitamins. The metabolic defect in these cases may be due in part to a deficiency of methyl groups. B vitamin supplementation supports the need for enzyme cofactors but cannot provide substrate in the form of methyl groups. L-Methionine is an essential amino acid and is required for protein synthesis, but it also plays a unique role in metabolism as *S*-adenosylmethionine, which is the primary methyl donor in metabolism. The observation that L-homocysteine, which is produced in the metabolism of L-methionine, is remethylated 2–4 times before it is destroyed is key to understanding the possibility of a methyl group deficiency. This suggests that the requirement for methyl groups (ie, *S*-adenosylmethionine) may be 2–4 times that for methionine in support of protein synthesis. L-Homocysteine can be remethylated to form L-methionine by betaine or N^5-methyltetrahydrofolate. Betaine and one-carbon sources that lead to the production of N^5-methyltetrahydrofolate and the remethylation of L-homocysteine to form L-methionine should be considered along with B vitamin supplementation in the treatment of homocystinuria and neural tube defects.

Key Words: Betaine • methyltetrahydrofolate • homocystinuria • neural tube defects

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