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Predictors of Survival in Children with Methymalonic Acidemia with Homocystinuria

We read with interest the recent report by Qiliang, *et al*. [1] on the outcome of 45 children diagnosed with combined methymalonic academia (MMA) and homocystinemia. The authors report a 40% mortality in their cohort. Apart from mortality, it would be important to know the degree and pattern of neuromorbidity in the survivors. Combined MMA and homocystinemia is a potentially treatable inborn error of metabolism, and high mortality and possibly high morbidity in the reported cohort need a careful evaluation. The high mortality in the reported cohort can partly be explained by the inadequate parenteral B_{12} replacement given to the patients. Highlighting the importance of meticulous long-term treatment, we wish to point out certain important aspects of management of children with combined MMA and homocystinemia.

The critical component of the treatment of combined MMA and homocystinemia is parentral B_{12}. This therapy has to be given in the adequate doses daily and lifelong. Hydroxyl-cobalamine injections are the only form of B_{12} proven to be beneficial in patients with this disorder. It is recommended that hydroxyl-cobalamine be given daily intravenously, subcutaneously or intramuscularly. The recommended dose of parenteral hydroxyl-cobalamine is 0.3 mg/kg/ day, once a day. The suggested targeted plasma B-12 levels are ≥1,000,000 pg/mL [2]. Previous reports have shown that the progression of complications in patients with combined MMA and homocystinemia arise in part due to inadequate hydroxyl-cobalamine. There are several reports of marked clinical and neurological deterioration in patients weaned from daily to less frequent dosing [3,4]. Hence, it is essential for all involved in the care of affected individuals to ensure daily administration of the injection. The monitoring parameters include serum MMA and total homocystine levels and normalization of plasma methionine and hematological parameters. The other co-factors recommended for use include oral Betaine (250 mg/kg/ day in 3 divided doses), oral Folinic acid (5-15 mg/day in 2-3 Divided doses), and oral Levocarnitine (50-100 mg/ kg/day in 3 divided doses).

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Editor's notes: *The corresponding author of the original paper referred to in this correspondence did not provide any response.*