



87.5% based on weight-for-age. Their PEM was marasmus evidenced by absence of both edema and serum albumin of less than 35 g/L. However, 75% had serum transferrin level of less than 2 g/L and the mean value was significantly lower than that of healthy children. Their serum transferrin levels are partly regulated by their iron store evident by significantly negative correlation between serum transferrin and ferritin levels. Children with Hb H disease did not show any significant differences in serum lipid and apoprotein levels from healthy children, whereas children with  $\beta$ -thalassemia/Hb E disease without and with splenectomy had significantly lower serum total cholesterol, HDL-C, phospholipid and apoprotein A-I levels than healthy children. These changes are most likely due to the derangement of hepatic function caused by iron overload and chronic hypoxia. All of the 3 groups of thalassemic children had significantly higher erythrocyte cholesterol and phospholipid contents than healthy children: 1.3-1.8 and 1.3-1.7 folds of those in healthy children. The increase in these erythrocyte lipid contents could be associated with the increase in the erythrocyte membrane surface area. Regarding to essential fatty acid status, biochemical linoleate depletion was detected in children with  $\beta$ -thalassemia/Hb E disease without and with splenectomy evidenced by their

significantly lower serum linoleic level than that in healthy children. Thus the effect of soybean-based formula supplementation was assessed in 18 children with  $\beta$ -thalassemia/Hb E disease postsplenectomy. Each thalassemic children participated in a 14-day supplementation of a 40g Gen-Formula, a cholesterol-free and iron-free, once daily, which provided 180 kcal and 6.8g of protein in addition to their habitual intake. Though there was significant increase in energy and protein intake in thalassemic children during receiving soybean based formula supplementation but no significant improvement in their height-for-age and weight-for age. However, the benefit of Gen-Formula supplementation on their visceral protein status was evident by the significant increase in serum RBP level at wk2. The mean total fat, linoleate and alpha-linolenate intakes in children with  $\beta$ -thalassemia/Hb E disease postsplenectomy were 27.0, 6.1, and 0.79% of total energy intake at wk0 and 28.1, 7.4, and 0.97% of total energy intake at wk2, respectively. Their linoleate and alpha-linolenate intakes at wk2 were significantly higher than those at wk0. Biochemical assessment also revealed that their serum and erythrocyte 18:2 n-6 levels at wk2 increased significantly and the values were not significantly different from those in healthy children. The result

indicate higher linoleate requirement in children with  $\beta$ -thalassemia/Hb E disease than the RDA for linoleate intake of 3% of total energy. It appears that linoleate intake of 7.4% of total energy intake is adequate to normalize serum and erythrocyte 18:2 n-6 levels in children with  $\beta$ -thalassemia/Hb E disease postsplenectomy. Gen-Formula supplementation for 2 wk did not alter the derangement of n-3 and n-6 polyunsaturated fatty acid metabolism in total erythrocyte lipids caused by iron overload. This is evident by no significant changes in erythrocyte 20:4 n-6, 20:5 n-3 and 22:6 n-3 levels. The study has also shown that courses of protein-energy malnutrition in thalassemic children were at least due to 3 factors: chronic hypoxia, iron overload, and inadequate protein-energy intake.