BACKGROUND: The ketogenic diet (KD) is a nutritional therapy for the treatment of epilepsies and certain metabolic disorders like the glucose-transporter-deficiency or the pyruvate-dehydrogenase-deficiency. The basis of the ketogenic diet is the change of energy metabolism to utilisation of fatty acids and their metabolites, ketonic bodies. Carnitines, which play an important role in transport and elimination of fatty acids, are essential for effective ketogenesis. Carnitine deficiency is described on ketogenic diet. The aim of this study is to evaluate when to expect a carnitine-deficiency during KD. PATIENTS: The carnitine levels of 19 patients aged 1.4 to 8.5 years (median 4.0 years), who were treated with ketogenic diet because of pharmacoresistant epilepsy, were evaluated retrospectively. RESULTS: Carnitine deficiency during KD was detected in 26 % of the patients and in 57 % of the patients without carnitine substitution. Decreased carnitine level occurred also with carnitine substitution independent from additional valproat therapy. The time of appearance of carnitine deficiency on KD was between 3 days and 248 weeks (median 32 weeks). CONCLUSION: Regular controls of carnitine levels should be performed during the treatment with ketogenic diet, both at the beginning and during longterm-therapy.