P-018 - COLOMBIAN PATIENTS WITH NEUROMETABOLIC/NEUROGENETIC DISEASES TREATED WITH KETOGENIC THERAPIES

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INTRODUCTION: Neurometabolic/neurogenetic diseases are a complex area in pediatric neurology with heterogeneous manifestations like ataxia, mental retardation and refractory epilepsy, because of these symptoms the use of ketogenic therapies (KT) is useful and is supported by the knowledge of its mechanisms of action and neuroprotective properties. OBJECTIVE: Evaluate the efficacy of KT in patients with neurometabolic/neurogenetic diseases attended by Metabolic Therapies (MT) in Colombia. METHODS: Multicentric retrospective observational descriptive study was carried out; We reviewed the data of patients with neurometabolic/neurogenetic diseases with RE, who received treatment with a KT with a formula based on medium chain triglycerides “KetoVOLVE®” as the sole source of food or combined with diet; during March 2014 to June 2018, throughout Colombia. Software SPSS v.22 was used. RESULTS: n= 19 patients, 53% male, median of age 4,8y (0,8y to 23,2y). Diagnoses: non ketotic hyperglycinemia (13; 68%), 2 hydroxy glutaric aciduria L (2; 10,5%), neuronal ceroid lipofuscinosis (2; 10,5%), glut-1 deficit (1; 5,5%) and deficit of 5-phosphate ribose isomerase (1; 5,5%). All patients receive KT but in different modalities determined by the ketogenic ratio (KR) like this: KR 1: 1 (32%), KR 2: 1 (21%), 2.5: 1 (5%), 3: 1 (21%), 4: 1 (21%) by enteral route feeding for 68% of patients. Median of time in treatment is 25,7 months (1 to 50 months). Mean of seizures/day before treatment 18, at six months of treatment, 1 seizure/day (decrease 95%) and after treatment, median 0 crisis/day. Secondary effects of KT reports: Constipation (5%), diarrhea (5%), improvement of constipation with the KT (37%). There was no change in the number of anticonvulsant drugs used. About nutritional status according with BMI there is no changes (-0,7 to -0,8 before and after KT). CONCLUSIONS: Significant improvement was found in the number of epileptic seizures and the clinical condition of patients with the management of the KT with formula based on medium chain triglycerides in patient with neurometabolic /neurogenetic diseases with RE in Colombia.