

## P-016 - CURRENT STATUS OF KETOGENIC DIET THERAPY IN PATIENTS WITH GLUCOSE TRANSPORTER 1 DEFICIENCY SYNDROME (GLUT1DS) IN ARGENTINA

Armeno M<sup>1</sup>, Verini A<sup>1</sup>, Eiroa HD<sup>2</sup>

*Hospital Pediatria J.P.Garrahan. Buenos Aires. Argentina 1 Servicio Nutricion 2 Servicio Errores Congenitos Metabolismo*

**INTRODUCTION:** Glut-1 deficiency is a metabolic disease, and the KD (ketogenic diet) is considered the first-line therapy in this syndrome, since ketones use another transporter to enter to the CNS that can provide the brain with an alternative source of fuel, correcting the impaired brain energy metabolism. **AIM:** We conducted a survey on the ketogenic diet therapy in 14 patients diagnosed with glucose transporter protein-1 deficiency syndrome (GLUT-1 DS) to evaluate the efficacy of the treatment with the classic ketogenic or modified Atkins diet from the viewpoint of patient's families. **METHOD:** A 3-page survey was distributed to all attendees of a family-centered meeting for GLUT1DS in November 2017, held at Hospital Pediatria Garrahan. The surveys were completed by parents, collected anonymously, and information analyzed in a database. **RESULTS:** Surveys were received from 14 families of patients diagnosed with GLUT-1 DS, 4 patients were females and 10, males with a median age 7,8 years (3 to 18). The mean diet duration was 8 years (range: 5 month-16.5 years). The types of KD therapies used were the Classic KD (10) and the Modified atkins diet (4). The ratio more frequently used was 4: 1, and most patients (13 of 14) used to measure urine ketones twice a week. The time between the diagnosis and the initiation of the KD was 6 month on average. The outcome in those with early diagnosis and intervention was surprisingly good. All but one patient with the classic phenotype became seizure free after treatment with the classic ketogenic or modified Atkins diet. Acetazolamide was effective in one patient with paroxysmal exercise-induced dyskinesia. **CONCLUSIONS:** the families complain about the late diagnosis but they showed a high level of satisfaction with the efficacy of the KD therapy. Although the long-term prognosis in patients with GLUT-1 DS partly depends on the underlying genetics, our study supports the assumption that early initiation of treatment with a ketogenic diet may positively affect the outcome.