**INTRODUCTION:**
Ketogenic diet has been used as a first line treatment in patients with GLUT 1D for fueling brain in a long-term basis. Bone health in children that use KD has been a concern due to the acid load associated with the diet, deficient calcium and vitamin D intake, and chronic anti-epileptic drug (AED) use (1). Dual energy X-ray absorptiometry (DEXA) has been recommended as a gold standard for assessing bone mineral density (BMD) in children because of its availability, reproducibility, speed, low exposure to ionizing radiation, and robust pediatric reference data (2). However, other risk factors have to be considered for a thorough evaluation such as chronic immobilization, low body weight, use of drugs that affect bone metabolism, hormonal abnormalities, personal and familial history of fractures (2,4). In the pediatric age group, the diagnosis of osteoporosis requires bone density measurement (BMD) levels ranging between 1.5 and 3.7 mmol/l (3, 4, 5, 6). At the time of DEXA, all patients had normal activity. Four subjects were overweight, 4 had normal weight and 2 were underweight at the time of DEXA. Just 1 subject was classified as short stature with H/A ≤ 2 SD.

**MATERIALS AND METHODS:**
Cross-sectional study. We conducted a retrospective review of clinical records of 11 patients (6 males), who followed a KD as a treatment for GLUT 1D in Santiago de Chile. Children where between 5-19y of age. DEXA (GE Healthcare; Lunar iDXA) was performed as part of our follow-up program for assessing bone status, by means of areal bone mineral density (g/cm²); results were adjusted for age and ethnicity. AEDs (40 cases) KD ratio, micronutrient intake and supplementation were collected from clinical and dietary record.

Anthropometric measures were registered according to (6, 22) score SD, including height for age (HA) and body mass index (BMI).

**Low for age BMDz** was defined with Z scores between -1.0 and -2.5 SD.

**BONE STATUS IN PATIENTS WITH GLUT 1 DEFICIENCY SYNDROME (GLUT1D) TREATED WITH KETOGENIC DIET (KD): A CHILEAN COHORT**

**RESULTS:**
Four patients were on classic KD (3, 4, 5, 6), and 6 patients were on a MAD diet with ketogenic ratios between 2.2-5.1 (Table 1). Beta-hydroxybutyrate (BHB) levels ranged between 1.5 and 3.7 mmol/l (3, 4, 5, 6). At the time of DEXA, all patients but 1 had normal activity. Four subjects were overweight, 4 had normal weight and 2 were underweight at the time of DEXA. Just 1 subject was classified as short stature with H/A ≤ 2 SD.

**DISCUSSION:**
The preservation of adequate bone health in patients on long term KD has been demonstrated as a difficult task, mostly in patients with neurological impairment.

In this cohort with GLUT 1D patients, BMDz measured by DEXA was used for bone status assessment. Even though half of the subjects presented with BMDz values below the norm, no subject had clinically significant morbidity. This could be explained because the risk of bone fragility is not only dependent on BMDz values, but also on age on onset and severity of the underlying condition and the number of associated risk factors (such as poor nutrition or inactivity). (2, 3) Patients had a KD ratio of 4.2 and all those patients had low BMDz. Those patients had a KD ratio of 4.2 and all those patients had low BMDz.

In this study, the majority of subjects had an adequate nutritional status, and only 2 of them had a low BMI, both of them having low for age BMDz along with one of them being non-ambulatory.

The effect of mechanical loading and exercise on bone accretion has been described previously (4); with exercise intervention leading to 0.6%-1.7% greater annual increase in bone accrual in patients with neurological impairment. Further research (8) demonstrated that non-walkers had lower mean BMDz that walkers in a cohort of 31 patients with cerebral palsy.

Further analysis demonstrated that there were no differences in BMDz regarding duration of KD and type of diet. This could be associated with adequate calcium and vitamin D consumption, which was prevalent in our cohort. Moreover, the BOH levels were alongside a conservative range (1.5-3.7 mmol/l).

Interestingly, the group with higher KD ratio (≥2.5:1) had significantly greater vitamin D levels. In our group vitamin D supplementation is not universal, but target according to vitamin D dietary intake, considering all GLUT 1D patients as entitled to fortificated vitamin D formula as part of a supplementary food soop Program. Intake varies among them according to preference for cooking OR not. This result could reflect that a greater intake of fats in the higher KD ratio diet has a direct impact on vitamin D intake. This finding could be significant for patients with poor adherence to pharmacologic supplementation, in whom we can use dietary sources to improve vitamin D levels and should be further explored.

Finally, bone health is a very important long-term issue in patients with GLUT 1D, treated with KD and a thorough evaluation should be included as part of our protocol for monitoring adverse effects. The evaluation considers DEXA but also assessment of nutritional status, physical activity, pubertal stage, ambulation, patient and family fracture history, and long-term AED exposure, which is not a regular practice in some centers, as demonstrated by Fong et al (9), who reported 84% of neurologists infrequently performed bone health screening investigations routinely.