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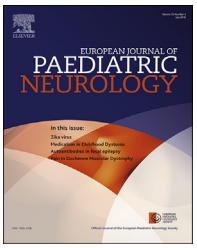

Original article

Questionnaire survey on the current status of ketogenic diet therapy in patients with glucose transporter 1 deficiency syndrome (GLUT1DS) in Japan

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ABSTRACT

Objectives: We conducted a questionnaire survey on the efficacy and side effects of ketogenic diet (KD) therapy in patients with glucose transporter 1 deficiency syndrome (GLUT1DS) as well as issues associated with long-term KD therapy from the viewpoint of patients' families.

Subjects and methods: The subjects were 34 patients whose ages at the time of the survey ranged between 2 and 50 years (median, 11 years). The ages at the diagnosis ranged between 3 months and 48 years and 5 months (median, 4 years and 10 months), and KD therapy was started within 5 months in all patients.

Results: The types of KD therapies used were modified Atkins diet (MAD) in 18 patients (53%), MCT (medium chain triglyceride)-KD in 9 (26%), classic KD in 5 (15%), LGIT (low-glycemic index treatment) in 1 (3%), and unspecified diet in 1 (3%). Epileptic seizures improved by more than 90% in 17 patients, by 50–89% in 9, by less than 50% in 3, and an unknown percentage in 5. Neurological symptoms other than the epileptic seizures improved markedly, moderately, and mildly in 14, 5, and 7 patients, respectively, and did not improve in 2. The side effects of KD therapy were seen in 9 patients and it was subsequently discontinued in one.

Conclusions: The families of patients showed a high level of satisfaction with the efficacy of KD therapy for the neurological symptoms. However, in order to continue KD therapy for a long period of time, its tolerability needs to be improved.

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Abbreviations: GLUT1DS, glucose transporter 1 deficiency syndrome; MAD, modified Atkins diet; MCT-KD, medium chain triglyceride-ketogenic diet; LGIT, low-glycemic index treatment.

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1. Introduction

Glucose transporter type 1 deficiency syndrome (GLUT1DS) is a metabolic encephalopathy caused by a defect in the transport of glucose into the brain, where it is the main substrate for energy metabolism.[1–3] Ketogenic diet (KD) therapy has long been considered to be an effective therapy for this disorder.[4,5] Since early KD therapy can reportedly prevent the sequelae of chronic cerebral glycopenia, the guideline of making an early diagnosis of GLUT1DS and early introduction of KD therapy have been recommended.[2,6] KD therapy was originally developed as a treatment for epileptic seizures in the 1930s, and was widely used to treat refractory epilepsy in children until the 1980s, when it was less recognized as a formal treatment choice owing to the advent of newly-introduced antiepileptic drugs. However, this diet therapy became the focus of attention again in the United States and Europe in the 1990s, and has recently been used worldwide to treat patients with refractory epilepsy.[7] There have been many ketogenic diet centers that provide nutrition guidance of KD for families set up not only in the United States and Europe, but also in Korea and the Middle East.[8] Due to the long history of KD therapy, its advantages and disadvantages have been studied in detail.[9] However, Japanese individuals eat a rice-based diet, and difficulties have been reported for Japanese patients to strictly maintain a diet high in fat and extremely low in carbohydrates for a long period of time. Therefore, KD therapy received less attention from the 1980s onwards, and has been used only in some hospitals specialized for epilepsy. However, the introduction of an easier-to-eat modified Atkins diet (MAD) coupled with an increase in the number of patients with GLUT1DS, and also approval for health insurance coverage in April 2016 as an alternative epilepsy treatment method once again have aroused increasing interest on this diet therapy.[10,11] A recent nationwide survey by the Health, Labor and Welfare Ministry's research group found that a large number of domestic patients receive KD therapy. MAD, which is less restrictive and easier to maintain for a long period of time, has become the mainstay for the treatment of GLUT1DS in Japan.[5,11] However, the usefulness of KD therapy, the effects of its long-term use, and the burden on patients' families have not yet been clarified in detail. Therefore, we herein evaluated the efficacy and side effects of KD therapy as well as issues associated with its long-term KD use from the viewpoint of patients' families, in cooperation with the GLUT1DS Patients' Association.

2. Subjects and methods

Among patients and their families belonging to the GLUT1DS Patients' Association (GLUT1DS Kanzyakai), those who understood the intent of this questionnaire survey, and offered the assistance with it were included in the present study. Questionnaires containing 21 items were prepared. The contents of the questionnaire included: current age, gender, age at onset, age at diagnosis, age at KD diet therapy initiation, types of KD therapies, KD therapy initiated at hospital or at home, urine ketone levels checked by ketone strips, efficacy for epileptic seizures, improvements in other neurological symptoms, side effects of KD therapy, current medication, and ingenuities and difficulties with the continuation of KD therapy. The efficacy for epilepsy seizures was defined as seizure freedom, >90% seizure reduction, 50–89% seizure reduction and <50% seizure reduction. We asked patients to subjectively evaluate and categorize improvements in neurological symptoms other than epileptic seizures into marked, moderate, mild, or no improvement.

After the approval of this study was obtained from the GLUT1DS Patients' Association, surveys were distributed to the attendees of the 2016 GLUT1DS networking event that were hosted by the association, and completed questionnaires were later received by mail.

In statistical analyses, the chi-squared test was performed using Excel software (BellCurve for Excel: Social Survey Research Information Co., Ltd.). A P-value <0.05 was considered to be significant.

3. Results

Questionnaires were sent to 46 members of the GLUT1DS Patients' Association. Thirty-four patients or their families completed and returned the surveys for analysis (Responder rate 74%). They included 21 female and 12 male patients and gender was not reported in the remaining one patient. Ages at the time of the survey ranged between 2 and 50 years (median, 11 years) (Fig. 1). Ages at the diagnosis of GLUT1DS were distributed between 3 months and 48 years and 5 months (median, 4 years and 10 months), and ages at the initiation of KD therapy ranged from 3 months to 48 years and 5 months (median, 5 years and 0 months) (Fig. 1). The duration from the diagnosis of GLUT1DS to KD therapy initiation ranged from 0 to 5 months (mean, less than 4 weeks). The mean duration of KD therapy was 69 months (range, between 4 and 168 months) (Fig. 2).

3.1. *The types of KD therapies used*

Eighteen patients (53%) received MAD therapy, 9 (26%) MCT (medium chain triglyceride)-KD therapy, 5 (15%) classic KD diet therapy, and 1 (3%) LGIT (low-glycemic index treatment). The type of KD was not reported in the remaining one patient. Among the 3 out of the 18 patients who received MAD therapy, 2 were switched from classic KD to MAD therapy, and 1 was switched from MCT-KD to MAD therapy. Conversely, 2 patients were switched from MAD to MCT-KD therapy because MAD therapy did not achieve any significant improvements. The details of MAD were described in 5 patients: carbohydrates were restricted to 12.5 g/day in 1 patient, 15 g/day in 1, 20 g/day in 2, and 45 g/day in 1. Among 9 patients treated by MCT-KD therapy, the ratios were 3:1 (n = 1), 2:1 (n = 3), 1.5:1 (n = 3) and 1:1 (n = 2). Four patients using classic KD therapy reported the following ratios: 2:1 in 3 and 1.5:1 in 1. KD therapy was started during admission in 30 patients and at home in 3 patients (MCT-KD therapy in 2 and classic KD therapy in 1). The remaining one patient did not report.

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