

THE CURRENT USE OF ALTERNATIVE DIETARY PROTOCOLS FOR GLYCOGEN STORAGE DISEASE TYPE III AMONG ITALIAN DIETITIANS

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ABSTRACT – Objective: To investigate the use of Alternative Dietary Protocols (ADPs) for treating Glycogen Storage Disease Type III (GSD III) by Italian dietitians and nutritionists working in Inherited Metabolic Diseases (IMDs).

Materials and Methods: A 16-question online survey was distributed to all Italian members of the Nutrition and Dietetic working group of the Italian Society for the Study of Hereditary Metabolic Diseases and Newborn Screening (SIMMESN), addressing ADPs usage, specific protocols, special medical foods, medium-chain triglyceride supplements, hydration, vitamins, and minerals. The survey also inquired about side effects, night-time meals, cornstarch use and emergency protocols.

Results: All dietitians/nutritionists reported using at least one ADPs for both adult and paediatric patients. The most common protocol was a high-fat, high-protein diet (66%), followed by the Modified Atkins Diet (MAD) (33%). Less frequently, ketogenic or high-fat diets were implemented. These dietary changes extended fasting intervals and reduced or eliminated night-time meals and cornstarch use without any reported hypoglycemic events.

Conclusions: This is the first study in Italy to examine the use of ADPs for GSD III. Further research is needed to provide clinical evidence on the efficacy of these dietary strategies in managing GSD III.

KEYWORDS: Glycogen storage diseases type III, Dietary practices, Ketogenic dietary therapies, Alternative dietary protocols, Atkins diet, High-protein, Low-carbohydrate.

LIST OF ABBREVIATIONS: ADP: Alternative Dietary Protocol; FSMP: Foods for Special Medical Purposes; GSD III: Glycogen Storage Disease Type III. CKD: Classic Ketogenic Diet; IMSs: Inherited Metabolic Diseases; LGIT: Low Glycemic Index Treatment; MAD: Modified Atkins Diet; MCT: Medium-Chain Triglyceride; SIMMESN: Italian Society for the Study of Hereditary Metabolic Diseases and Newborn Screening.

INTRODUCTION

Glycogen Storage Disease type III (GSD III) (#MIM 232400) is an autosomal recessive disorder of glycogenolysis caused by a deficiency of the debranching enzyme. This enzyme deficiency leads to impaired glycogen breakdown, resulting in reduced glucose availability and hypoglycemia, as well as intracellular accumulation of abnormal glycogen with short outer chains (limit dextrin) in various tissues, primarily the liver, muscles, and heart. The key biochemical features of GSD III include hypoglycemia (with or without ketosis), hyperlipidemia, and elevated liver transaminases^{1,2}.

The primary clinical manifestations are fasting intolerance, ketotic hypoglycemia, hepatomegaly, and failure to thrive^{1,3}. Over time, patients may develop skeletal myopathy with muscle fatigability and cardiomyopathy, potentially leading to serious complications such as liver adenomas, cirrhosis and hepatocellular carcinoma^{2,4}. Hypertrophic cardiomyopathy may present in early childhood. GSD III-related endocrine disorders include type 2 diabetes and osteopenia^{5,6}, and females with GSD III are at an increased risk of polycystic ovary syndrome⁷. The main goals in managing GSD III are to maintain normoglycemia, prevent secondary metabolic complications, and mitigate long-term adverse outcomes.

The primary strategy for GSD III management is dietary intervention. Traditional recommendations include a high-carbohydrate diet, increased protein intake (20–25% of total energy in cases with cardiomyopathy), restricted fasting intervals, uncooked cornstarch supplementation, and continuous overnight feedings to prevent hypoglycemia^{4,8}. Carbohydrates from food and cornstarch, along with amino acids from proteins, support euglycemia by acting as glucose sources and substrates for gluconeogenesis⁴.

Although traditional dietary interventions can prevent fasting hypoglycemia, they do not prevent cardiomyopathy, which can still develop and prove fatal during long-term follow-up⁹. However, various Alternative Dietary Protocols (ADPs) focusing on dietary lipid modification have shown positive effects on cardiomyopathy and muscle function in GSD III. These ADPs are high-fat, high-protein diets with a low carbohydrate intake, calculated as percentages of macronutrients which may or may not induce a state of ketosis¹⁰⁻¹³.

The Modified Atkins Diet (MAD) is a ketogenic diet which involves a carbohydrate intake of 10-20 g/day, with or without Medium-Chain Triglyceride (MCT) supplementation¹⁴⁻¹⁶, while other Classic Ketogenic Diets (CKDs) promote ketone production through fixed macronutrient ratios, are primarily used to treat drug-resistant epilepsy^{9,17,18}.

Given the limited studies on heterogeneous populations of children and adults with varying severity of GSD III, the role of ADPs and their specific lipid and carbohydrate adjustments have

not been systematically evaluated. Additionally, current guidelines lack clear recommendations for ADPs implementation, resulting in inconsistent application of these protocols across countries, including Italy, potentially affecting patient outcomes^{4,19}. This study aims to examine the current use of ADPs in Italian dietitians/nutritionists, exploring the associated challenges and opportunities for managing GSD III.

MATERIALS AND METHODS

In September 2023, a 16-question multiple-choice survey ([Supplementary Material](#)) was distributed online to 19 Italian dietitians and nutritionists of the Nutrition and Dietetic Working Group of SIMMESN through their respective clinical center contacts. The survey aimed to investigate the current use of ADPs in the management of GSD III.

The questionnaire covered the following topics (multiple responses were allowed for all questions):

1. The use of ADPs for patients with GSD III;
2. The specific ADPs applied include CKDs, the MAD, high-fat diets, high-fat and high-protein diets and the low glycemic index treatment (LGIT). Respondents were also asked to specify the ketogenic ratio;
3. The use of foods for special medical purposes (FSMPs);
4. The use of MCT supplements and FSMPs containing MCT fats;
5. Water intake and supplementation with vitamins and minerals;
6. Side effects and locations where diet changes are implemented;
7. Management of night-time meals and cornstarch use;
8. The use of emergency protocols, including emergency letters.

Ethical approval was not required, as the survey did not include clinical outcomes or patient-specific data.

Statistical Analysis

The responses were analyzed using descriptive statistics, with data presented as percentages (%) and absolute numbers (n). As a multiple-choice survey, participants could report one or more responses; therefore, the sum of percentages for each item does not correspond necessarily to 100.

RESULTS

All dietitians/nutritionists responded to the survey for a total of 19 responses received; 63% (12/19) reported treating patients with GSD III. Among these, 50% (6/12) treated both pediatric and adult patients, 33% (4/12) treated only adults, and 17% (2/12) treated only children. All participants managing patients with GSD III reported using one or more ADPs, in agreement with the center clinician. Specifically, 67% (8/12) used a high-fat and high-protein diet, followed by MAD in 33% (4/12) and CKDs in 17% (2/12). Additionally, 8% (1/12) reported using the LGIT, a high-fat diet, or MCT supplementation (Figure 1).

Ketonemia monitoring was also explored, as part of the strategy for clinical management. While 42% (5/12) of respondents did not measure ketones, 58% (7/12) considered an optimal ketonemia range to be between 1 and 3 mmol/L. None regarded levels >3 mmol/L as ideal. These ketonemia levels refer to both children and adults.

In terms of applying a ketogenic ratio within dietary protocols, respondents reported to use one or more dietary approaches: 33% (4/12) used a 1:1 ratio, 25% (3/12) used a 2:1 ratio, and 17% (2/12) used a 3:1 ratio. None of the participants applied a 4:1 ketogenic ratio. Half of the respondents (6/12) calculated the percentage of lipids rather than using a ketogenic ratio.

Regarding the use of FSMPs, 67% (8/12) of respondents incorporated them into their dietary protocols. Pasta and rice were used by 75% (6/8), followed by fixed-ratio ketogenic beverages (2:1, 3:1, 4:1) in 63% (5/8) of cases. Bread and substitutes were also used by 63% (5/8). Biscuits and other sweet foods were included by 50% (4/8), while pudding and protein supplements were used by 25% (2/8).

The use of MCT fats was common, with 50% (6/12) of dietitians using them as supplements and 42% (5/12) providing them through FSMPs containing MCT fats.

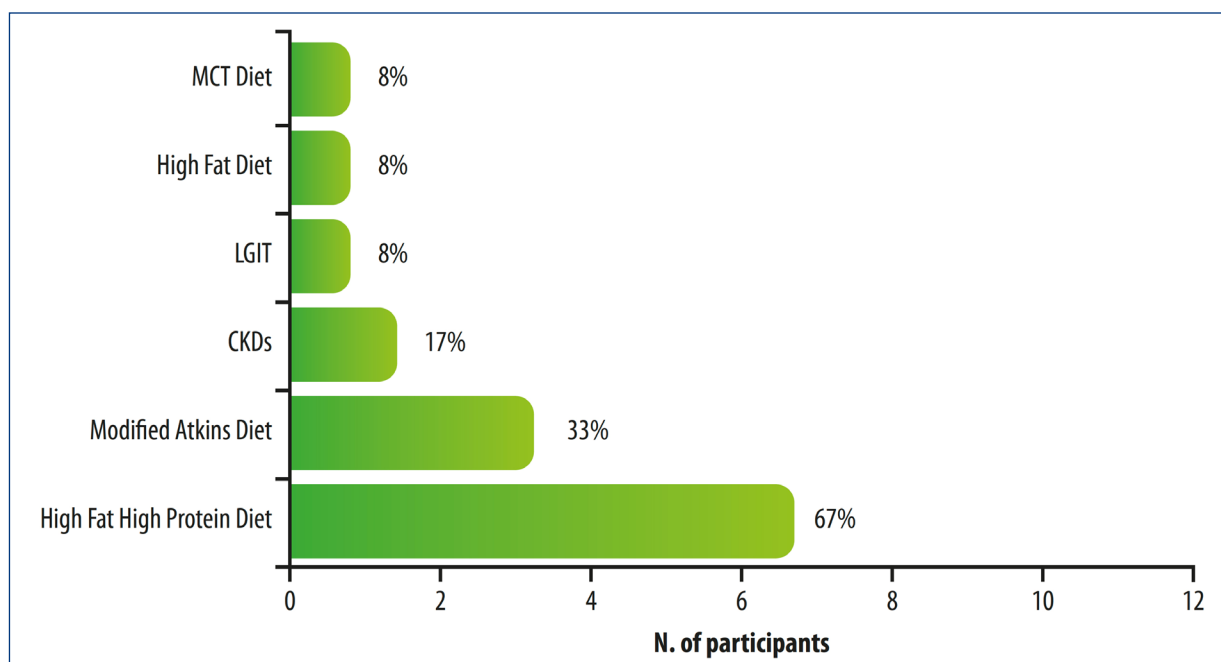


Figure 1. Utilization of ADPs protocols among Italian Dietitians following GSD III patients.

The survey also investigated the use of other supplements. Multivitamins and vitamin D were used by 58% (7/12) of respondents, while potassium citrate was used by 25% (3/12). Docosahexaenoic acid was reported by 17% (2/12), and calcium and lactic acid bacteria probiotics were used when needed by 8% (1/12). No respondents reported the use of fiber, magnesium, or carnitine. Additionally, 92% (11/12) of participants recommended or prescribed water intake as part of the dietary protocol.

Potential adverse effects associated with ADPs were also evaluated. Aside from cases of worsening osteoporosis, constipation, nausea, and headache, reported by one dietitians/nutritionists each, no significant side effects – including hypoglycemia, loss of appetite, weight loss, or kidney stones – were reported (75%, 9/12) (Figure 2). Regarding the location setting for initiating a dietary protocol switch, 67% (8/12) of respondents indicated that the transition occurred at home, 8% (1/12) during hospital admission, and 25% (3/12) reported that both options were possible.

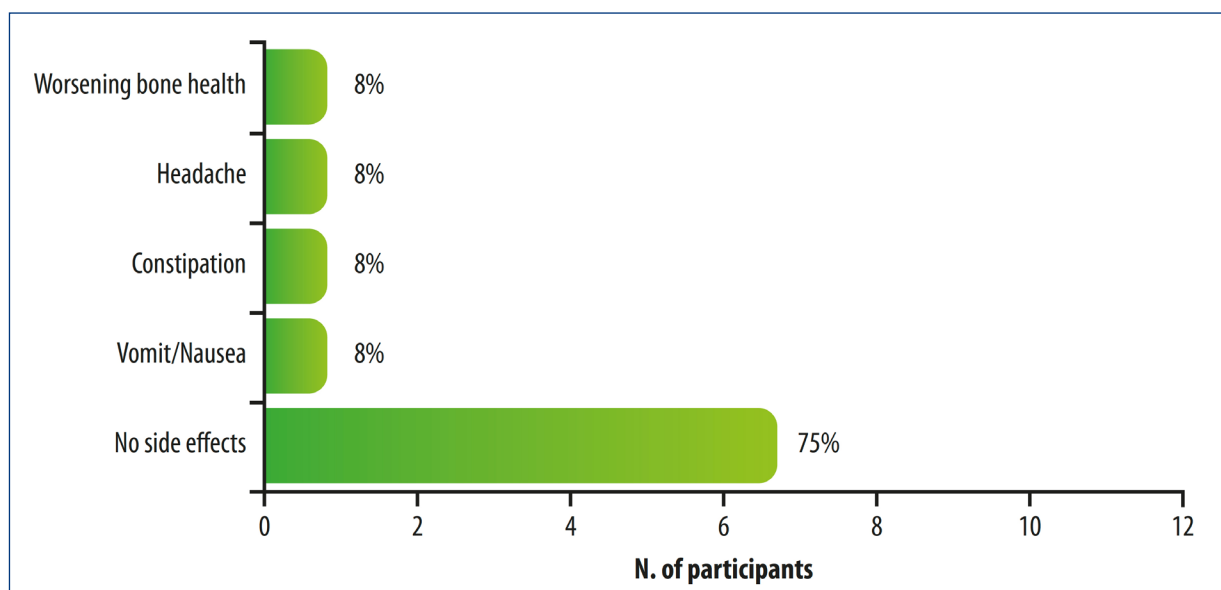


Figure 2. N° of Italian Dietitians reporting specific side effects of ADPs on GSD III patients. No hypoglycemia was observed.

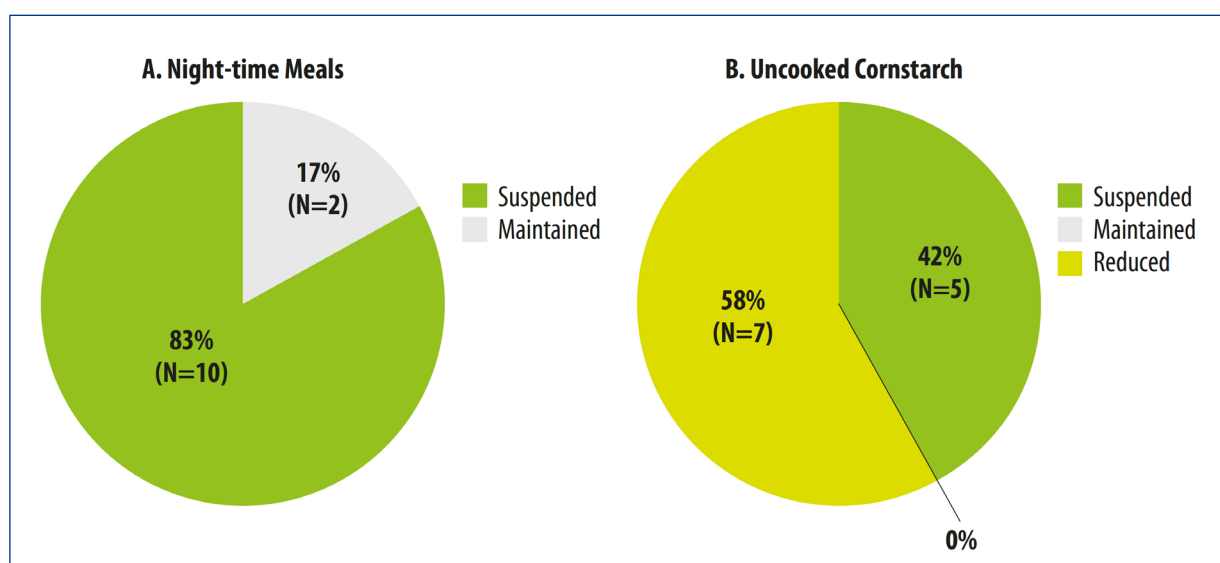


Figure 3. Management of night-time meals (A) and uncooked cornstarch (B) after ADPs introduction among the dietitians.

Night meals were eliminated, or fasting intervals were extended in 83% (10/12) of cases. Cornstarch was removed from the diet in 58% (7/12) of cases and reduced in the remaining 42% (5/12) (Figure 3). Furthermore, 42% of participants reported not issuing a new emergency letter for patients following alternative treatments compared to traditional approaches.

DISCUSSION

We conducted a nationwide survey among all Italian members of the Nutrition and Dietetic working group of SIMMESN to investigate the current use of ADPs for the management of GSD III and to identify any differences or gaps in the applied approaches. All participants reported using at least one ADPs for patients with GSD III, having moved away from traditional protocols in agreement with the center clinician. This indicates a consensus among Italian dietitians regarding the appropriateness of ADPs, given the effectiveness of these dietary practices in improving cardiac and muscular outcomes, reported in literature. Indeed, both adult and pediatric patients with GSD III have shown benefits from ADPs, with improvements in the energetic state of the heart and skeletal muscles, leading to better biochemical and clinical outcomes and, ultimately, enhanced quality of life^{14,18}. Furthermore, ADPs have been shown to improve clinical symptoms, biochemical markers, and echocardiographic findings, particularly in cardiomyopathy, and have led to a reduction in transaminase and creatine kinase levels, stabilization of blood glucose, prevention of unwanted weight loss, and a slowing of disease progression^{10,12,16,17}.

In half of the cases (50%; 6/12), ADPs were applied to a heterogeneous population of both children and adults with varying degrees of impairment, necessitating different dietary treatments. The most commonly used protocol was the high-fat, high-protein diet (66%), followed by MAD (33%). Less frequently, dietitians/nutritionists reported the use of CKDs, MCT supplementation, the LGIT, or high-fat diets. As previously reported in literature concerning pediatric epilepsy^{20,21}, the MAD protocol appears to be a more flexible and tolerable option than CKDs, facilitating long-term adherence. Similarly, the high-fat, high-protein diet is characterized by specific macronutrient percentages rather than a fixed ketogenic ratio, which may explain its preference in long-term GSD III management¹⁰⁻¹².

Regarding the measurement of ketones, 42% (5/12) of dietitians/nutritionists reported not measuring ketones, while 58% (7/12) considered moderate ketonemia (1–3 mmol/L) to be optimal. This finding could have several explanations. First, most centers may prefer using moderate-lipid diets, such as high-fat, high-protein protocols, the Modified Atkins Diet (MAD), or low-ratio Ketogenic Diets (CKDs). These approaches typically do not result in consistent ketone production throughout the day or may lead to lower levels of ketonemia.

Second, respondents may not consider ketone production to be a critical goal for the clinical management of patients with GSDIII, and for this reason, they may opt for dietary approaches with moderate lipid content.

Finally, the absence of specific guidelines might lead respondents to select dietary approaches solely based on the individual needs of the patient.

Other practical aspects of dietary management, such as the prescription of FSMPs, MCT fats, water intake, and supplements, were also addressed. In 67% (8/12) of cases, FSMPs were provided free of charge. The most commonly used FSMPs were pasta, rice, bread/sandwiches, and ketogenic beverages, specific products with fixed ketogenic ratios (1:1 to 4:1). These products, also used in macronutrient-based ADPs without a fixed ketogenic ratio, facilitate meal preparation and improve adherence to dietary guidelines. Pasta and bread substitutes are particularly useful for patients transitioning from traditional diets, which previously included higher carbohydrate intake.

MCT fats, available in oil or powder form, were used as supplements by 50% (6/12) of dietitians/nutritionists, while 42% (5/12) reported using FSMPs containing MCT fats, such as cookies, creams, and beverages. In literature, MCT fats are known to be advantageous due to their higher ketogenic potential compared to long-chain fatty acids²², making them useful in the dietary management of disorders involving carbohydrate metabolism²³.

Hydration and vitamin supplementation were routinely recommended by most participants, with multivitamins and vitamin D prescribed by 58% (7/12) of respondents. This is particularly important given the carbohydrate restrictions of ADPs, which limit the intake of fruits, vegetables, and grains. Proper hydration and supplementation are known to be crucial to prevent kidney stones and ensure adequate micronutrient intake, as seen with CKDs²¹.

A notable finding was the low incidence of side effects reported by 75% (9/12) of dietitians/nutritionists. Apart from isolated cases of worsening osteoporosis, constipation, nausea, and headache, no significant adverse effects – such as hypoglycemia, loss of appetite, weight loss, or kidney stones – were observed. This supports the safety of ADPs in clinical practice. Furthermore, 67% (8/12) of dietitians/nutritionists implemented the dietary switch at home rather than in a clinical setting. Despite the elimination or reduction of cornstarch and night meals in many cases, no hypoglycemic events were reported. Indeed, these protocols are known to help reduce glycogen overload in the heart and muscles while maintaining euglycemia by activating gluconeogenesis from neo-gluconeogenic amino acids and promoting energy production from the oxidation of alternative energy substrates to carbohydrates, such as fatty acids and ketone bodies²⁴.

Moreover, 58% (7/12) of participants had not yet developed a specific emergency protocol for patients on ADPs. In a retrospective, observational, single-center study, Rossi et al²⁵ reported on the generation of a website www.emergencyprotocol.net, designed to support families and health-care providers to generate personalized emergency letters for patients with hepatic GSD and the main FAOD. However, given the recent implementation of these protocols, further refinements are likely needed.

Limitations

This study has some limitations. Biochemical and organ function data from patients with GSD III were not collected. Including such data, alongside the survey results, would provide a more comprehensive understanding of the long-term impact of ADPs on clinical outcomes, such as cardiomyopathy, liver dysfunction, growth, and dyslipidemia. A multicenter observational study involving clinicians and including both biochemical and dietary data from homogeneous patient groups is necessary to evaluate the long-term efficacy of ADPs in GSD III management. Furthermore, as the time of ADPs treatment for patients with GSD III was not investigated in this survey, the absence of diet-related complications due to a short follow-up cannot be excluded completely.

Despite these limitations, this study represents the first Italian investigation into the use of ADPs for GSD III and offers valuable insights for future research. We believe that continued discussions among dietitians and nutritionists from the SIMMESN Dietetics and Nutrition Group will contribute to the harmonization of guidelines and practices, ultimately improving dietary management and defining a specific protocol for GSD III.

CONCLUSIONS

In this study, we conducted the first nationwide investigation in Italy on the application of various ADPs for the management of GSD III by all members of the Nutrition and Dietetic working group of SIMMESN. All participants reported using at least one ADP despite the absence of standardized guidelines. Following the dietary switch, longer fasting periods, suspension of night meals, and reduction or complete elimination of cornstarch were achieved without any reported hypoglycemia events. Further research is needed to provide systematic clinical evidence supporting the promising potential of ADPs in the management of patients with GSD III.

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All data generated or analyzed during this study are included in this published article and its [supplementary material](#).

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The authors declare that they have no conflict of interest to disclose.

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