The efficacy of alternative treatments in maintaining metabolic control of Glycogen Storage Disease (GSD) Type I

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Glycogen storage disease (GSD) is a rare genetic disorder that disrupts proper metabolic function with consequences of hypoglycemia, hyperlactatemia, hyperuricemia, and hyperlipidemia that ultimately compromises health. Maintaining stable blood glucose levels through starch therapy is the current standard treatment. However, such onerous therapy requires frequent feedings and interrupts the normal sleep cycle of patients and their families. The purpose of this systematic review was to evaluate the efficacy of alternative treatments in comparison to the standard starch therapy for maintaining metabolic control in patients diagnosed with GSD Type I. A MEDLINE and EBSCO search was conducted using search terms “glycogen storage disease” and “treatment.” Studies were limited to those that addressed GSD Type I, were written in English, and were published between 1990 and 2014. Of the six studies reviewed, two evaluated the effectiveness of a modified experimental starch, one evaluated the use of a medium-chain triglyceride supplement, one evaluated dextrose and a combination of dextrose and cornstarch, and two evaluated aggressive cornstarch therapies. Changes in levels of blood glucose and lactate levels were reported or described in all studies as indicators of metabolic control. Overall, this research indicates the importance of maintaining metabolic control in the treatment of GSD Type I through stable blood glucose levels and lowered lactate levels. Due to the diverse needs of each patient, standard treatments are difficult to establish. Future research is needed to evaluate the effectiveness of novel treatments for GSD among a wider subject population for sustained time periods.