Metabolic Control, Quality of Life, and Body Image in Patients with Glycogen Storage Disease Type Ia

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Glycogen storage disease is a group of inborn errors of metabolism, with type Ia being the most common form of the disorder. Glycogen storage disease type Ia (GSDIa) is a multisystemic condition in which individuals have various complications secondary to an inability to properly break down glycogen and to perform gluconeogenesis. Complex management is then necessary for patients and includes dietary modification, frequent cornstarch usage, and evaluation for additional complications such as hepatic adenomas, hypertriglyceridemia, and kidney disease. Previous studies have found lower scores in quality of life and body image in GSDIa patients; however, the specific factors influencing this relationship remain unknown. In this study, 24 adult participants (n=24) with glycogen storage disease type Ia completed a survey including measures of health-related quality of life, body image, and metabolic control. Results found that quality of life was significantly lower than the general population on both the physical and mental component scores ($t=-3.11, p=0.005; t=-2.21, p=0.03$). Additionally, body image was significantly lower on all subscales: Weight ($t=-5.88, p<0.001$), Appearance ($t=-5.67, p<0.001$), and Attribution ($t=-2.38, p=0.02$). In general, significance was not reached when examining roles that certain metabolic and demographic factors play in health-related quality of life and body image. Therefore, the relationship between these factors is most likely complex. Overall, the current study confirms previous findings of lower health-related quality of life and body image in this population and provides preliminary evidence on potential factors influencing this phenomenon.

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