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RESEARCH REPORT

Disordered Eating and Body Esteem Among Individuals with Glycogen Storage Disease

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Abstract Glycogen storage disease (GSD) is an inherited disorder that requires a complex medical regimen to maintain appropriate metabolic control. Previous research has suggested the disease is associated with decreased quality of life, and clinical experience suggests that patients are at risk for disordered eating behaviors that may significantly compromise their health. The current study assessed eating attitudes, eating disorder symptoms, and body image among 64 patients with GSD ranging from 7–52 years old ($M = 18.5$ years old). About half the participants were male ($n = 33$, 51.6%). Most participants were diagnosed with GSD Type I ($n = 52$, 81.3%). Quantitative and qualitative analyses were utilized. Results indicated that 14.8% of children and 11.1% of adolescents/adults with GSD met the clinical cutoff for dysfunctional attitudes toward eating, suggesting high likelihood for presence of an eating disorder. However, traditional eating disorder symptoms (e.g., binging, purging, fasting, etc.) were less prevalent in the GSD sample compared to population norms ($t = -6.45$, $p < 0.001$). Body esteem was generally lower for both children and adolescents/adults with GSD compared to population norms. These results were consistent with interview responses indicating that GSD patients experience negative feedback from peers regarding their bodies, especially during childhood and adolescence. However, they reported growing acceptance of their bodies with age and reported less negative attitudes and behaviors. Assessing mental health, including symptoms of disordered eating and low body esteem, among individuals with GSD should be an important component of clinical care.

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Introduction

Glycogen storage disease (GSD) is a complicated disease with a complex medical regimen. For many patients, the recommended treatment includes restricted intake of non-glucose sugars, including lactose, fructose, and sucrose (Rake et al 2002a, b), and individuals with GSD Ia may require supplemental cornstarch every 4–5 h around the clock (Weinstein and Wolfsdorf 2002). This rigid feeding schedule can interfere with daily activities and interrupt sleep schedules of patients and their families. Patients may also require additional daily dietary supplements and medications to combat associated morbidities (Chen 2011).

Relatively little research has examined the mental health impact of living with GSD. One notable study of GSD Ia and Ib patients found that they reported decreased quality of life (QOL) related to physical health, psychosocial health, and social functioning compared to the general population (Storch et al. 2008). Yet, it remains unknown what aspect(s) of GSD might negatively impact well-being. One hypothesis is that GSD may negatively influence self-esteem, which is an important predictor of life satisfaction and mental health (Diener and Diener 1993). Body esteem, a particular domain of self-esteem, is of particular interest in the GSD population. Characterized as the self-evaluation of physical appearance, body esteem has been shown to be lower in

overweight and obese individuals (Mendelson et al. 1996; O'Dea 2006). For individuals with GSD not under optimal metabolic control, hepatomegaly can lead to abdominal distention and increased BMI (Özen 2007). Additionally, the regular consumption of uncooked cornstarch as part of treatment necessitates a large and sometimes excessive caloric intake. This is compounded in some patients by exercise intolerance due to myopathy (Kollberg et al. 2007), restrictions on allowable exercise (Mundy et al. 2005), as well as the need to compensate for increased activity with additional cornstarch therapy (Shin 2006). Certain forms of GSD also present with physical manifestations including poor growth, short stature, distended abdomen, and thin extremities (Wolfsdorf et al. 1999). Body dissatisfaction and low self-esteem are considered risk factors for the development of disordered eating (Furnham et al. 2010; Olmsted et al. 2008).

Importantly, clinical experience suggests that many patients with GSD exhibit some form of disordered eating. Given how crucial dietary compliance is for patients with GSD, the serious risks of maladaptive eating behaviors can have even more detrimental consequences. Assessing for low body esteem and addressing contributing factors may provide insight into ways to improve quality of life for GSD patients and prevent further complications. This study examined body esteem, eating attitudes, and disordered eating in patients with GSD. Specifically, we hypothesized that 1) patients with GSD would demonstrate lower levels of body esteem, more dysfunctional eating attitudes, and higher rates of disordered eating than population norms, and 2) patients with more severe forms of GSD (i.e., Types Ia and Ib) would demonstrate lower levels of body esteem, more dysfunctional eating attitudes, and higher rates of disordered eating than patients with less severe forms.

Methods

Participants

Participants were 64 individuals with glycogen storage disease who were being followed by the University of Florida GSD Program. All participants had a diagnosis of GSD based upon demonstration of abnormal enzyme activity on a liver biopsy or genetic mutation analysis. The sample included 33 male participants (51.6%) and 31 female participants. They ranged in age from 7 to 52 years old ($M = 18.5$). Half the sample ($n = 32$) were children aged 7–12; the others were 13 years or older. The majority of participants ($n = 52$, 81.3%) were GSD Types Ia or Ib. Other represented types included Type IIIa (6.3%), Type VI (1.6%), Type IX (7.8%), and unclassified (3.1%).

Procedure

All procedures were approved by the University of Florida Institutional Review Board. All GSD patients ages 7 and older were invited to participate in this study during their scheduled medical monitoring. A trained research assistant obtained informed consent/assent from patients and/or their parents. Next, participants completed a semi-structured interview and a number of age-appropriate self-report psychological assessment questionnaires. Family members and medical staff were asked to leave the room in order to allow honest responding. Participants were reminded at regular intervals that their families and clinical care team would not have access to their responses.

Measures

Eating Disorders Inventory-3 (EDI-3)

This self-report inventory is useful in assessing individuals suspected of having an eating disorder (Garner 2004). The 91 items are rated on a 6-point Likert-type scale. A sample item is "I exaggerate or magnify the importance of weight." The EDI-3 has demonstrated strong psychometric properties.

Eating Disorders Inventory-Child (EDI-C)

The EDI-C is a multidimensional self-report questionnaire that is specially designed for children and young adolescents (Garner 1991). Based on the EDI-2, it consists of 91 items, scored on a 6-point Likert-type scale. Sample items include "I think my stomach is too big" and "I am very afraid of getting fat." The EDI-C has demonstrated good reliability and validity.

Eating Attitudes Test (EAT)

The EAT is used to assess eating attitudes among adults (Garner and Garfinkel 1979). The 26 items, rated on a 6-point Likert-type scale, include "I eat diet foods" and "I display self-control around food." The EAT yields scores on 3 subscales: Dieting, Bulimia and Food Preoccupation, and Oral Control. Using the cutoff score of 20, it has been used to identify individuals with any DSM-IV-defined eating disorder with a 90% accuracy rate, though it does not predict specific diagnoses (Mintz and O'Halloran 2000).

Children's Eating Attitude Test (ChEAT)

The ChEAT is a 26-item version of the Eating Attitudes Test that is used to assess eating attitudes that are common

in youth with eating disorders (Maloney et al. 1988). Items are scored on a 6-point Likert-type scale using the guidelines developed by Anton et al. (2006). Sample items include "[I] feel guilty after eating" and "[I] exercise to burn energy." The ChEAT yields subscale scores for Body Concern, Dieting, Food Preoccupation, Weight Gain, Vomiting, and Calories.

Body Esteem Scale (BES)

The BES is used to identify dissatisfaction with one's body (Mendelson et al. 2001). Adapted from the Body Esteem Scale for Children (BES-C; see below), the 30 items are rated on a 5-point scale with three subscales: Appearance (general feelings about their own appearance including face, hair, etc.), Weight (satisfaction specifically related to body size and weight), and Attribution (beliefs about how others evaluate the respondent's appearance). Sample items include "I like what I see when I look in the mirror" and "I feel I weigh the right amount for my height."

Body Esteem Scale for Children (BES-C)

The BES-C is a reliable and valid measure of body dissatisfaction for children (Mendelson and White 1985). It consists of 20 items rated on a 4-point Likert-type scale, which measure Appearance, Weight, and Attribution. Respondents rate statements such as "There are lots of thing I'd change about my looks if I could." Due to low reliability ratings for the Attribution subscale, only the Appearance and Weight subscales are recommended for interpretation (Mendelson et al. 1996).

Open-Ended Interview

All subjects participated in a short semi-structured interview about their experiences living with GSD. One of the questions was, "Everyone feels differently about their bodies. Tell me a little about how you feel about your body." Optional prompts included, "What are the things you like about your body? What are the things you don't like about your body?".

Data Analysis

Descriptive statistics were computed for study variables. Questionnaire scores were transformed into *z*-scores to allow for comparisons across age groups. One-sample t-tests compared GSD patient scores to population norms. Independent samples t-tests compared GSD Type I patient scores to scores for other types. In order to be sensitive to factors that may be affected by normal development, data were generally analyzed separately for children (ages 7–12) versus adolescents/adults (13 and older). Semi-structured interview responses were transcribed from digital audio recordings. The grounded theory method was used to assess important themes (Corbin and Strauss 2008).

Results

On measures of traditional eating disorder symptoms (EDI-3/EDI-C), GSD patients overall scored lower than the published population norms ($t = -6.45$, $p < 0.001$). Patients with more severe forms of GSD (i.e., Type Ia or Ib) demonstrated a trend toward lower levels of eating disorder symptoms than patients with less severe forms ($t = 1.77$, $p = 0.08$).

With regard to eating attitudes, 14.8% of the children scored above the cutoff score of 20 on the ChEAT ($M = 13.29$; SD = 6.56), indicating dysfunctional attitudes toward eating and likely presence of an eating disorder. Although the ChEAT does not yield specific diagnostic information for particular disorders, these results indicate clinically significant symptoms. Average scores were highest for the Dieting subscale among children in this sample. Similarly, 11.1% of the adolescents/adults scored above the cutoff score of 20 on the EAT-26 ($M = 12.8$; SD = 6.48), suggesting clinically significant symptoms. Average scores on the 3 subscales were not significantly different. There were no significant differences between male and female patients with GSD on either the ChEAT or EAT.

On measures of body esteem, GSD patients generally scored lower than population norms, indicating less healthy beliefs. Specifically, although scores for children with GSD were not significantly lower for body esteem related to their overall Appearance ($t = -1.07$, *ns*), they reported lower body esteem related to Weight ($t = -3.01$, $p = 0.006$). In addition, adolescents/adults with GSD reported lower body esteem related to others' perceptions of their bodies (Attribution: $t = -2.86$, $p < 0.007$) and trends toward lower body esteem for overall Appearance ($t = -1.90$, $p < 0.07$) and Weight ($t = -1.82$, $p < 0.08$). When comparing standardized scores that were calculated based on gender norms, male and female patients with GSD did not differ in their self-reported body esteem scores.

The sample BMI ranged from 15.3–39.9. In total, 49.2% of the sample had a "healthy" BMI between 18.5 and 25.0, whereas 38.1% had BMI in overweight/obese category. Among the children, BMI was unrelated to scores on the ChEAT or total score on the EDI-C. Higher BMI was positively correlated with scores on the "Body Dissatisfaction" subscale of the EDI-C ($r = 0.52$, $p = 0.006$) and also correlated with lower body esteem for Weight ($r = -0.42$, $p < 0.04$) and overall Appearance ($r = -0.36$, $p < 0.07$). In the adolescent/adult group, BMI was not significantly

related to scores on the EAT. However, BMI positively correlated with eating disorder symptoms on the EDI-3 total score ($r = 0.54$, $p = 0.002$), as well as the "Drive for Thinness" subscale ($r = 0.46$, $p = 0.10$) and the "Body Dissatisfaction" subscale ($r = 0.60$, $p = 0.001$). Higher BMI also correlated with lower body esteem for Weight ($r = -0.68$, $p < 0.001$) and overall Appearance ($r = -0.44$, $p < 0.02$). Though results demonstrated that adolescent/adult height ranged from 3′11″ to 5′8″ [$M = 5'3''$ (SD = 4.3″)], height was not related to body esteem. The association between height and body esteem was not analyzed in participants under 13.

From the semi-structured interviews, six major themes associated with body esteem arose, including (1) issues with bullying/teasing, (2) weight concerns, (3) height concerns, (4) positive body image, (5) negative body image, and (6) age-related acceptance of GSD and its effects on the body.

Bullying/Teasing

Some younger patients and several adult patients reported bullying and teasing while in school. Weight and appearance were particular topics of concern. For example, one girl mentioned that other kids at school "are really skinny and it is kind of embarrassing not being able to be like that... Sometimes they will call you fat," while a young woman acknowledged that "it wasn't easy dealing with weight issues when I was younger... I used to get kind of made fun of and ... called fat." Others indicated bullying occurred because of GSD-related differences from peers. One male teenager remarked, "I got a lot of [flak] in school for actually doing the routine, and taking the medicine, and not eating the same stuff they were," while a middle-aged woman noted, "growing up I was always mocked because I had a big liver and I was clumsy and bad in sports."

Weight

Weight concern was a common theme across age and gender. For some, it appeared to be entirely self-perpetuated [e.g., "I think I'm ... a little overweight and don't know why I feel like that" (female child) and "I think that I'm fat, but everyone says I'm not" (male child)]. However, several patients were particularly concerned with how they believed others perceived them. One young woman remarked, "I wish people knew I had GSD, that I don't just eat everything in sight, and thus am overweight ... because I work out four times a week, and I try to eat well and I'm still on the much much higher side. I know if people saw me they would just come to the conclusion that obviously I must not eat right." Another exclaimed, "I hate and I will cry every time... I am asked if I am pregnant... I hate that my body looks like this and it's not my fault because it is this genetic disease" (young woman). Of concern, a few patients specifically related their weight concerns to their cornstarch treatment, saying, "I'm getting like 1,500 calories from cornstarch alone right now, and no matter how much I exercise I will never be a certain amount of shape I want (sic) ... and there is absolutely nothing I can do about it" (male teenager). One young woman even admitted to dangerously reducing her treatment, saying, "I wish I could be thinner ... like any other women. I got really thin last summer ... because I reduced my cornstarch more than I should have."

Height

Though often ameliorated by appropriately aggressive treatment and management, short stature is a common complication of the glycogen storage diseases. Surprisingly, several patients viewed their height as a positive attribute. A young woman noted, "I was always teased about being short, but it made me special and people gave me attention. I love attention." Similarly, an adolescent girl reported, "I like being short." However, other participants viewed their height negatively or expressed a desire to be taller. For example, a middle-aged woman stated, "Wish I was taller." A young man noted that his height affected participation in group sports saying, "If I played with my peers, since I was so much smaller, I would get dominated. So what they did is that they ... put me in a group setting that was a year or two years younger, so I could be more competitive."

Negative Body Image

When asked what they did not like about their bodies, patients' responses ranged from overall dissatisfaction [e.g., "I don't like my body ... a problem since I was a little kid. I really wish I didn't have this disease" (middle-aged woman), and "I have long been at odds with my body because it would never do what I wanted it to do... Even when I was older I still always thought I was fat and then now, it's like failing ... the muscles are failing" (middle-aged woman)] to more specific complaints. For example, several individuals commented on the GSD side effect of having large livers/stomachs [e.g., "I guess you always feel self-conscious because you have the tendency to have the larger liver, which makes your stomach larger" (young woman), and "When I was younger, I had a little pot belly and I resented that, but that's behind me now" (young man)]. Other GSD-related issues were also mentioned, including "I have a scar from my liver biopsy. I don't like that!" (young woman). One patient expressed frustration with her thin limbs, "Fractures are frustrating. I have spindly arms and legs. I wish they were more muscular"

(middle-aged woman). Another young man who displayed typical Type Ia physiology expressed, "I don't like my gut. I mean I've got skinny little arms. My nickname's the Tick with everyone I know." Notably, none of the patients identified common body concerns that were unrelated to GSD (e.g., amount/type of hair, acne, size/shape of nose, feet, breasts, etc.)

Positive Body Image

A few respondents did report positive attributes of their bodies, including GSD treatment-related improvements. One young girl said, "I'm really proud about my body because . . . my liver is really healthy, and I breathe really well, and I think really well, so I'm really proud of that." Another expressed happiness with her overall appearance saying, "My core body I actually feel pretty good about . . . I feel I am a pretty attractive person, cute . . . I'm pretty tall for a girl of GSD, sort of above my age . . . when I'm fit, I actually feel pretty darn good about myself" (young woman).

Age-Related Acceptance

Many of the adult participants spoke about age-related acceptance of their condition and reported becoming more comfortable with their appearance in adulthood. As one young man explained, "Now that I'm older, I don't really care. But when I was in my teenager years I didn't like my big belly, I was self-conscious to go to the pool, take my shirt off. . . Now that I'm old, it's normal to be out of shape so it doesn't bother me that much. . ." Another commented, "With GSD, you know most people aren't a double zero. . . I did have a hard time with that in high school. . . I've had a baby now. So I mean, anytime I have any sort of bodily issue, it's like, well, you know, I've had a baby. And that to me is more important than squeezing into a double zero" (young woman)]. Finally, another young adult woman remarked, "as you get older, you really accept the way you are."

Discussion

In this study, GSD patients reported fewer classic symptoms of eating disorders (e.g., binging, purging, fasting, etc.) than population norms, with Type I patients demonstrating fewer symptoms than patients with less severe forms. However, the GSD patients also reported elevated dysfunctional attitudes toward food and eating, as well as lower body esteem. These results are concerning, as both increase the risk for disordered eating (Mintz and O'Halloran 2000; Littleton and Ollendick 2003; Verplanken and Velsvik 2008).

Unhealthy habits may be adopted by patients with GSD because healthy weight management options are limited due to dietary requirements and restrictions on exercise, particularly for patients with GSD Type I (Mundy et al. 2005). The current results suggest that a significant minority (11–15%) of GSD patients have clinically significant disordered eating symptoms and may meet diagnostic criteria for Eating Disorder Not Otherwise Specified. Given how crucial food is as medicine for GSD patients, even subclinical eating disorders pose a severe threat for this population. It is possible that traditional measures of eating disorder symptoms are not appropriate for GSD patients, given that the items primarily assess symptoms that could have catastrophic or even fatal consequences for GSD patients (e.g., fasting, purging, excessive exercise, etc.). As a result, GSD-specific measures may be needed to fully examine the extent of disordered eating and dysfunctional attitudes in this population. Items should assess behaviors such as skipping cornstarch doses, decreasing cornstarch without medical supervision, eating foods that are not well absorbed to manage weight, and unhealthy exercise habits.

In this sample, negative attitudes related to eating and body image appeared to be particularly problematic for younger patients, with higher average scores on the ChEAT and increased frequency of scoring above the clinical cutoff for children with GSD compared to non-clinical samples. Scores in the adolescent/adult group of GSD patients were more similar to body esteem scores in the general population, though the GSD sample still reported lower body esteem (Maloney et al. 1988; Maloney et al. 1989). These findings are consistent with the qualitative descriptions provided by adult GSD patients in this study, who reported more negative views during childhood, with improvements to their body esteem as they entered adulthood.

Associations between BMI and body esteem in the current sample were similar to those seen in a study of healthy young adults, where correlations between BMI and Weight Esteem ranged from $r = -0.43$ to -0.58 ($p < 0.05$), based on gender, and BMI and Appearance Esteem correlated $r = -0.36$ ($p < 0.05$) for both men and women (Streeter et al. 2012). A study of healthy children (Bernier et al. 2010) also demonstrated similar correlations between BMI percentile and overall body esteem ($r = -0.36$ to -0.45, $p < 0.01$); however, those children demonstrated a negative correlation between BMI percentile and scores on the ChEAT ($r = -0.22$ to -28, $p < 0.01$). When comparing to patients with chronic metabolic disease, previous research involving patients with Type I diabetes has noted that body dissatisfaction is associated with poor glycemic control, whereas good metabolic control is linked to better quality of life (Meltzer et al. 2001; Hoey et al. 2001). Poor metabolic control in GSD leads to greater complications and comorbid factors including hypoglycemic episodes, hepatomegaly, hyperlipidemia, hyperlactatemia, nephropathy, and hepatic adeno-

mas, which may negatively impact quality of life. Future research should explore links between metabolic control, body image, and quality of life, particularly in GSD patients suffering from overtreatment obesity.

The results of the present study should be interpreted within the context of some important limitations. First, though the sample was relatively large considering GSD is a rare disease, the small sample size likely affected statistical significance, particularly with regard to exploration of gender or age discrepancies. Second, the sample was overwhelmingly comprised of GSD Type I patients, precluding meaningful analyses among the subtypes. Third, the sample was comprised of individuals who attend regular clinic appointments, suggesting higher rates of compliance. This may affect their psychological response to living with GSD, including the burdens of the intensive medical regimen and the likelihood of fewer medical complications, resulting in somewhat biased results. Fourth, there are concerns about the appropriateness of certain study measures (i.e., measures assessing classic eating disorder symptoms) for this particular population. Fifth, the self-report nature of the data may have introduced response bias. Despite these limitations, this study is the first to examine the important topic of disordered eating and body esteem among patients with GSD.

Conclusions

Focus on food as essential medicine in patients with GSD may be protective against development of classic eating disorder symptoms, even in the presence of dysfunctional eating attitudes and low body esteem. The GSD patients in this study generally reported lower rates of eating disorder symptoms such as binging, purging, and fasting, with Type I patients, who have the strictest dietary guidelines and increased reliance on cornstarch therapy, reporting the fewest symptoms. Despite this, some patients reported concerning symptoms, and food preoccupation may have a negative impact on body esteem and quality of life for patients with GSD. Screening for dysfunctional eating attitudes, disordered eating behaviors, and negative body esteem should be included in the standard of care for patients with GSD. Questionnaires assessing the particular concerns of this population, as have been developed for individuals with diabetes, are needed to better understand their level of risk. Negative body image appears to be problematic in this population, and the medical team should assess and address this with young patients, in particular. Referral to qualified mental health professionals may be indicated for patients who are struggling with these concerns. At minimum, the medical team should provide patients with age-appropriate suggestions on how to talk with others about their disease and its impact on their body in order to reduce feelings of shame and decrease the potential for bullying or discrimination.

Compliance with Ethics Guidelines

Synopsis

Patients with Glycogen Storage Disease are at risk for disordered eating behaviors and low body esteem; these areas should be evaluated as part of standard clinical care.

Conflict of Interest

All the authors of this chapter declare that there are no conflicts of interest.

Informed Consent

All procedures followed were in accordance with the ethical standards of the responsible committee on human experimentation (institutional and national) and with the Helsinki Declaration of 1975, as revised in 2000 (5). Informed consent was obtained from all patients for being included in the study.

Author Contributions

Theresa B. Flanagan – Contributed to data analysis and the writing of the initial draft of the manuscript. Jill A. Sutton – Conducted statistical analysis and assisted with the write-up of results. Laurie M. Brown – Assisted with data collection and reviewed the final draft of the manuscript. David A. Weinstein – Provided the initial idea for the study and gave feedback on study design, elicited funding, and provided edits to the final manuscript. Lisa J. Merlo – Designed the study, supervised data collection, conducted statistical analysis, and contributed to the writing of the manuscript/guarantor.

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