A Novel Online Group Mindfulness Intervention for Hyperphagia in Prader-Willi Syndrome

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Introduction

Prader-Willi Syndrome (PWS) is a rare developmental disability that occurs in approximately one out of every 15,000 births resulting from genetic mutations on chromosome 15 (Dykens et al., 2007). The syndrome is generally characterized by mild to moderate intellectual disability, various social/cognitive deficits, and behavioral challenges including rigidity, temper outbursts, and hyperphagia—a lifelong overactive and insatiable appetite (Dykens et al., 2007). Hyperphagia is among the most disruptive and life-threatening challenges for individuals with PWS. Hyperphagia is comparable to constant starvation, resulting in extreme food-seeking behaviors such as hiding/stealing food or eating unpalatable items. Individuals with PWS are thus at risk, not only for obesity and obesity-related diseases such as cardiovascular disease, but also for premature death due to choking or stomach perforation as a result of large food intake (Dykens et al., 2007). Although clinical trials are underway, medical interventions such as bariatric surgery and weight loss drugs have not proven efficacious in this population (Mahmoud et al., 2023). Accordingly, families must often resort to food safety tactics such as locking cabinets and refrigerators to limit access to food. Maintaining these safety measures—among others—can cause conflict between individuals with PWS and their families and inhibit them from participating in social settings where food may be present (Dykens et al., 2007). This population, therefore, is in dire need of a novel intervention to manage hyperphagia.

Optimally, these interventions should be agency-respecting and individual-centered, rather than caregiver-centered. Mindfulness has produced promising results in other populations with intellectual and developmental disabilities but has yet to be sufficiently implemented and studied for managing hyperphagia in PWS (Singh et al., 2008). Through this project, I therefore hope to add to previous research findings on the efficacy of online group interventions in PWS, with the added dimension of using mindfulness techniques as a novel way to manage the severity of hyperphagic symptoms in PWS.

Literature Review

Hyperphagia is life-threatening in part because it leads to obesity and obesity-related health complications, including death. For this reason, PWS was previously conceptualized as a "genetic model of obesity" (Holland et al., 2003). However, in 2003, Holland et al. proposed a new way of thinking of this phenotype: individuals with PWS lack a satiety response while consuming food, and the body then misinterprets this state of not feeling "full" as one of hunger and/or starvation. It would, therefore, be more accurate to refer to PWS as a "genetic model of starvation" that frequently manifests as obesity in the context of our food culture, accessibility, and consumption patterns today (Holland et al., 2003).

With this concept of hyperphagia in mind, researchers have tried a variety of methods in an attempt to manage this behavioral phenotype. In a 2023 review, Mahmoud et al. synthesized the findings of several randomized clinical trials testing the efficacy of novel drugs or devices aimed at reducing hyperphagic symptoms in PWS. These include beloranib, oxytocin, setmelanotide, DCCR (Diazoxide Choline Controlled-Release), Livoletide, Cannabinoids, Exenatide, and transcranial direct-current stimulation (tDCS). Mahmoud et al. also comments on the use of surgical management, primarily bariatric surgery, as problematic in individuals with PWS due to postoperative complications. Mahmoud et al.'s review identifies two pharmaceutical weight-loss treatments—beloranib and DCCR tablets—that initially produced promising results in treating hyperphagia; however, clinical trials for the former were discontinued in

response to reported participant deaths, and trials for the latter require more time and a larger sample size to determine the true efficacy of the drug in PWS (Mahmoud et al., 2023).

In an attempt to better assess the effect of these interventions on hyperphagia in PWS, Dykens et al. developed a 13-item questionnaire to measure hyperphagic severity (2007). The questionnaire, named the Hyperphagia Questionnaire, measures the severity and type of various food-related preoccupations commonly observed in individuals with PWS. The questionnaire includes items asking about how clever/fast one's child is when obtaining food, the amount of time one's child spends talking about food, etc. One limitation of this questionnaire, however, is that it is designed to be completed by a parent or caregiver, not the individual with PWS themselves. The developers of the questionnaire chose this approach in response to previous attempts to measure hyperphagia, which required verbal responses from participants. Because individuals with intellectual disabilities often have difficulty accurately and thoroughly expressing their thoughts, the Hyperphagia Questionnaire was designed using a parent-report model in hopes of collecting accurate, reliable responses (Dykens et al., 2007).

This approach, however, is limited, as it bypasses the individual with PWS and instead transfers even more responsibility onto the caretakers of those with PWS who are often already overworked and overwhelmed (Kayadjanian et al., 2018). This is due to this population's unique, hyperphagia-related needs, and caretakers of individuals with PWS report higher stress levels, financial strain, and impact on quality of life when compared to the caretakers of individuals with other intellectual and developmental disabilities. In a 2018 study assessing caregiver burden in PWS, researchers used the Zarit Burden Interview (ZBI) to demonstrate that caregivers of individuals with PWS experience high levels of caregiver burden with an average ZBI score of 44.4 ± 15.4 with range 0 (no burden) to 88 (highest burden). This burden can take many forms, but high levels of caregiver burden were correlated with negative changes in mood, sleep, and romantic relationship satisfaction. The study also revealed that caregivers of teenage and young adult individuals with PWS face the highest levels of caregiver burden. Further studies are needed to determine the association between caregiver burden and caregiver-centered interventions in PWS. Regardless, these findings indicate the importance for future research in developing non-caregiver-centered interventions for hyperphagia in the PWS population (Kayadjanian et al., 2018).

The presence of intellectual disability in the PWS population may initially indicate the need for caregiver presence and participation during interventions. However, researchers in a 2022 study developed and administered a social skills intervention—named BOSS (Building Our Social Skills)—in which individuals with PWS joined groups of 6-8 PWS peers and worked together with two clinicians to improve their social cognition, motivation, and communication (Dykens et al., 2022). The intervention was conducted over a 10-week period and, upon analysis, produced significant improvements in the social cognition, motivation, and communication subscales of the Social Responsiveness Scale (Dykens et al., 2022). This study demonstrates the possibility of online, group-based, and individual-centered interventions, as opposed to caregiver-centered interventions for PWS. Although the study provides promising results for online social skills interventions in this population, it does so only for young individuals (M age = 20.8, SD = 6.42). It is not yet clear if this model can be used in interventions for older adults and for other behavioral phenotypes of PWS, such as hyperphagia.

The group-based, individual-centered model for intervention is important for several reasons, the first of which—a need to mitigate caregiver burden—has already been discussed. In addition, it is necessary from an ethical standpoint that individuals with PWS feel respected and able to participate in their own treatment and syndrome-management. This cultivates a sense of agency—which is often denied to those with intellectual and developmental disabilities. In a paper detailing stigma in mental health care, philosopher Heney (2022) discusses self-stigma—stigma that an individual places on themselves. Although PWS is different from mental illness in significant ways, PWS individuals are also prone to self-stigmatization in response to their hyperphagic behaviors. They may, as

a result, lie about their food-related behaviors and view hyperphagia as an aspect of their condition that is both undesirable and uncontrollable (Dykens et al., 2022). According to Heney, this type of pessimism results in a negative feedback loop, in which the individual is treated as a helpless agent, which further negatively impacts their ability to make improvements to their behavior (2022). For this reason, it is important that hyperphagia interventions treat individuals with PWS as active agents capable of making improvements in their own condition. This begins by including them via individual-centered interventions in their own symptom/behavioral management.

Some researchers may be skeptical about the willingness and ability of individuals with PWS to participate in their own interventions. A paper by Dykens et al. (2022), however, helps combat these concerns, treating PWS individuals as a rich source of insight for ways in which PWS treatments can be developed and adjusted. The study used semi-structured interviews—both individual and group—with the aim of better understanding the self-perceptions of people with PWS. Multiple themes emerged from this qualitative study. Theme 1 was "struggles with hunger and food seeking," with the following three subthemes: 1) "weight loss is hard," 2) "negative impacts on life choices, relationships," and 3) "strategies to manage hunger" (Dykens et al., 2022). It is evident from the quotations included in the study that the PWS participants 1) have a desire to manage this symptom, and 2) are able to participate, at least somewhat, in the management of their hyperphagia. For example, one participant—Thomas—remarks, "Like the food around me, it's like a drug that kills me. The food is a poisonous drug that kills me." Another participant—Douglas—states, "I am good at my own diet. I do well." These quotations demonstrate that at least some PWS individuals are aware of the severity of their condition, have a desire to manage their symptoms, and possess the capacity to try adhering to diet plans. To be certain, not everyone with PWS will share the willingness and/or ability to participate in an individual-centered intervention. However, this study approaches insights from individuals with PWS through a lens of curiosity and respect and, in doing so, indicates the possibility for individual-centered interventions in this population (Dykens et al., 2022).

A 2013 pilot study examined the effectiveness of a group mindfulness intervention for Williams Syndrome (WS)—a neurodevelopmental disorder resulting from an abnormality on chromosome 7 (Miodrag et al. 2013). Williams Syndrome also results in mild to moderate intellectual disability, as well as distinct internalizing and externalizing behaviors such as anxiety and impulsivity, respectively. The study used salivary cortisol and self-reported anxiety measures to assess the effectiveness of the intervention on participants with WS. The results indicated that salivary cortisol levels decreased on average after each mindfulness session, indicating not only that individuals with intellectual and developmental disabilities can participate in their own interventions but also that these interventions work. Although additional studies are needed, this pilot study provides preliminary evidence for the feasibility of group mindfulness interventions in populations with intellectual and developmental disabilities. (Miodrag et al., 2013).

Because hyperphagia has serious social, health, and behavioral consequences, it is important to accurately measure the prevalence and severity of hyperphagic behaviors. The current standard in the field is to administer the Hyperphagia Questionnaire, a 13-item questionnaire administered to caregivers of individuals with PWS to assess food-seeking behaviors (Dykens et al., 2007). The questionnaire includes questions such as, "How persistent is your child in asking or looking for food after being told 'no' or 'no more'?" and, "Outside of normal mealtimes, how much time does your child spend talking about food or engaged in food-related behaviors?" (Dykens et al., 2007). The Hyperphagia Questionnaire is integral to research on hyperphagia in PWS and allows researchers to measure the effect of behavioral and pharmacological interventions on this behavioral phenotype. However, in an attempt to return voice and agency to individuals with PWS, there is new research supporting the development of a self-report measure for hyperphagia in individuals with PWS (Dykens et al., forthcoming). This new self-report measure, named My Hunger Questionnaire, is a 22-item questionnaire administered to participants verbally and visually

using an online interview format (Dykens et al., forthcoming). This project will use both caregiver and self-report measure data to assess the efficacy of the intervention on hyperphagic behaviors.

In summary, there are several gaps in the current literature on the management of hyperphagia in Prader-Willi Syndrome. Firstly, there has still been relatively little success in managing hyperphagia as a behavioral phenotype, and many families must resort to trial and error in managing their child's extreme food-seeking behaviors (Dykens et al., 2022). This indicates that this population could benefit from a novel intervention to manage hyperphagic symptoms. Secondly, these interventions should be agency-respecting and therefore individual-centered, not caregiver-centered. Finally, online group interventions have produced promising results but have yet to be implemented to manage hyperphagic behaviors.

Methods

Participants

Sixteen U.S. adults between the ages of sixteen and thirty with genetically confirmed Prader-Willi Syndrome (PWS) will participate in the study. Participants will be recruited using the existing PWS research participant database provided by the Foundation for Prader-Willi Research. Individuals with PWS currently participating in other clinical trials related to PWS will be excluded from the study. Prior to the study, participants will be assessed via REDCap and virtual interview using the Hyperphagia Questionnaire (caregiver-report measure) and My Hunger Questionnaire (self-report measure), respectively. The study will be approved by the University's Institutional Review Board. Written informed consent will be obtained from parents and/or caregivers of participants with PWS. Individuals with PWS will also provide written assent before participating in the study.

Design

This longitudinal study explores the effectiveness of an online, group mindfulness intervention for hyperphagia in PWS. Due to the rarity of the syndrome, the intervention will take place online over Zoom to ensure that participants from a variety of locations can participate in the study. The study will employ widely-used and validated mindfulness techniques such as body scanning, as well as additional meditation and breathing techniques proven effective in other intellectual and developmental disability populations (Miodrag et al., 2013; Singh et al., 2008). The study will use the Hyperphagia Questionnaire and My Hunger Questionnaire scores to ensure stratified random assignment into either the treatment group or the control group, with each group consisting of two subgroups with four participants each. A structured group format will be used to ensure that 1) all participants receive the potential benefits of peer support and 2) members in each intervention group receive identical intervention methods. The peer support groups will run concurrently with the intervention groups to serve as a control. The control groups will receive the intervention at the conclusion of the study to ensure all participants reap the potential benefits of the intervention. All participants—including the control group—will be assessed at three points: 1) before the intervention/peer support group, 2) immediately following the completion of the intervention/peer support group, and 3) at a one-month follow-up.

Procedure

Both the intervention and control groups will meet for 30 minutes two times weekly over a period of five weeks during the summer. Meetings will take place at a mutually convenient time for each group of participants but will always occur at the same time to establish a consistent routine. A researcher assistant in the lab will be trained to administer and facilitate the meetings for both intervention and control groups. Time will be allocated for each session as follows: welcome activity (5 minutes), check-in/reflection (5 minutes), introduce and practice the new mindfulness technique (10 minutes), brainstorm session (5 minutes), and session reflection/check-out (5 minutes). The style and content of the check-in and mindfulness activities will mirror that which has been engaging and successful for younger audiences and other intellectual and developmental disability populations. The welcome activity will be used to build comfort and rapport among group members, and session check-ins will

be used to gauge participant mood and reflect on any techniques used between the last and current session. After learning the new mindfulness technique, participants will be asked to brainstorm specific times and ways to implement the new technique into their daily life. As a session check-out, participants will be asked to share brief feelings about the session, as well as any goals for the period between sessions.

The control group sessions will be similar in length and structure, except that no techniques or strategies will be introduced by the research assistant. Instead, participants will use the extra time to discuss shared interests, difficulties, or other PWS-related topics that interest them.

Expected Results and Discussion

The findings for this study will provide valuable insights into the efficacy and feasibility of an online mindfulness intervention for young adults with PWS. Based on the analyses discussed above, I expect that individuals who receive the mindfulness intervention will display a significant decrease in hyperphagic symptoms from baseline to one-month follow up as measured by the Hyperphagia Questionnaire and My Hunger Questionnaire. Although the study is underpowered, I predict that the 3 x 2 (time wave by group) repeated measures ANOVAs will demonstrate significant interactions between time and group for the intervention group. This would suggest that the intervention group observed a greater change compared to the control group, providing preliminary evidence on the effectiveness of the intervention.

Individuals with PWS need an effective intervention that aims to manage the severity of their hyperphagic behaviors. I predict this project will contribute to the current literature by providing evidence that supports feasibility and preliminary effectiveness of group mindfulness interventions in PWS. The findings will add to a growing body of evidence that individuals with PWS can actively participate in their own syndrome management when agency-respecting, individual-centered interventions are available. Unlike pharmacological or caregiver-centered interventions, mindfulness may offer a more accessible and empowering approach to managing hyperphagic symptoms.

Despite the prediction that the study will produce significant results, limitations include the small sample size, potential challenges navigating the online meeting format, and variability in cognitive and behavioral functioning of participants within each of the groups. Future studies should build on these results with larger, more diverse samples and explore additional outcomes related to overall well-being. Still, this study is an important step in expanding intervention options for hyperphagia in PWS through an individual-centered approach.

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About The Author

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