Received: 1 March 2021 | Revised: 9 August 2021 | Accepted: 30 September 2021

DOI: 10.1111/jar.12950

ORIGINAL ARTICLE

JARID Journal of Applied Research in Intellectual Disabilities bild WILEY

# 'The cure for us is a lot of things': How young people with Prader-Willi syndrome view themselves and future clinical trials

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## Abstract

**Background:** Despite work on the self-identities of people with intellectual disabilities, research has yet to describe the self-perceptions of people with Prader-Willi syndrome (PWS). The perspectives of those with PWS are also important for rapidly evolving clinical trials aimed at treating symptoms of PWS.

**Method:** Twenty-one young people with PWS were administered a semi-structured interview that assessed how they perceive their syndrome and clinical trials. Transcribed interviews were reliably coded using content-driven, applied thematic analyses.

**Results:** Five themes emerged: struggles with chronic hunger and food-seeking that impede goals and relationships; struggles with anxiety and outbursts, schedule changes and school; distancing from PWS; needs for clinical trials that cure PWS, reduce hunger or anxiety, and lead to improved outcomes; and needs for advocacy and awareness of PWS.

**Conclusions:** Findings shed new light on the self-perceptions of those with PWS and have important implications for current interventions and future clinical trials.

KEYWORDS
anxiety, clinical trials, cure, hyperphagia, Prader-Willi syndrome, self-perceptions

## 1 | INTRODUCTION

People with intellectual disabilities have unique insights into their lives and situations that are increasingly captured in qualitative research (Beail & Williams, 2014). Such work has, for example, identified the self-perceptions and social identities of people with intellectual disabilities (e.g., Beart, 2005; Kittelsaa, 2014; Logeswaran et al., 2019). From a disability rights perspective, this work gives voice to a group that has historically been marginalised and excluded.

Of concern, however, is that research has yet to probe the perspectives of people with specific causes of intellectual disability. Several studies have examined the self-perceptions of individuals with Down syndrome (e.g., Cunningham & Glenn, 2004), yet research is needed on the perceptions of those with other neurodevelopmental syndromes, including Prader-Willi syndrome (PWS).

A relatively rare disorder, PWS is caused by the lack of paternally derived imprinted genes to chromosomal region 5q11-q13, either through paternal deletions that vary in size, maternal uniparental disomy, or imprinting defects (see Cassidy & McCandless, 2010 for a review). People with PWS exhibit a complex phenotype that includes hyperphagia and heightened risks of obesity; mild to moderate intellectual disabilities; social cognitive deficits; compulsivity, rigidity and insistence on sameness; anxiety; and temper outbursts (e.g., Dimitropoulos et al., 2012; Dykens et al., 2017, 2019; Dykens & Roof, 2008; Whittington & Holland, 2018). Hyperphagia, often cast as the hallmark of PWS, onsets in childhood and is associated with dysfunctional neural networks involved in both satiety and reward. The impaired satiety response results in a state in which individuals are habitually hungry yet rarely feel full (Manning & Holland, 2015). Holland et al. (2003) have aptly described PWS as a state of starvation that manifests as obesity in food-rich environments.

Managing the life-threatening hyperphagia in PWS requires interventions that ensure food security (Cassidy & McCandless, 2010). The types or intensities of these interventions may differ across individuals but often include locking food sources (e.g., kitchen cabinets, pantries, refrigerators, garbage cans), vigilant supervision around food at home and in school or community settings, a reduced calorie diet (due to a low resting metabolic rate) and offering meals or snacks at predictable times.

While the severity of hyperphagia varies across individuals and over time, people with PWS often engage in food seeking behaviours, sometimes in very clever ways (Dykens et al., 2007). They may sneak food, manipulate others or bargain for food, and repeatedly ask about meals. Understandably, most individuals do not want to get into trouble for sneaking food, and are thus prone to lying, denying or distorting their food-related behaviours.

Although they may be unreliable reporters of food-seeking behaviours, people with PWS still have important perspectives to share about themselves. To date, only one study has explored how people with PWS view themselves. Using semi-structured interviews, Plesa-Skwerer et al. (2004) compared the self-concepts of young people with PWS versus Williams syndrome. Unlike those with Williams syndrome, participants with PWS reported feeling more marginalised and burdened by their condition. They further described how challenges with food and weight impeded their social lives and independence.

Beyond their self-perceptions, work is also needed on how people with PWS understand new treatments for their condition. PWS is now attracting considerable interest from pharmaceutical companies. Clinical trials testing the efficacy of novel agents in curbing hyperphagia and related symptoms have been completed, and others are currently underway or planned for the future (Dykens et al., 2018; Griggs et al., 2015; Miller et al., 2015).

As these trials evolve, however, the voices and perspectives of those living with PWS have not been heard. What might those with PWS seek in new medicines? What are their perceptions of the impact of such treatments?

In these mixed-methods, primarily qualitative study, 21 adolescents or young adults with PWS were administered semi-structured interviews that explored: (1) how they understand their syndrome and its affects, and (2) their opinions regarding new treatments for PWS. Interviews were coded using a content driven, applied thematic analytic approach. Secondarily, we explored possible associations between coded interview data and cognitive functioning, gender and other demographics.

## 2 | METHODS

### 2.1 | Participants

The sample included 21 home-living individuals with PWS aged 14–38 years ($M$ age = 20.71; SD = 7.70). They resided in the United States, 57% were females and 52% were still in high school. See Table 1 for other participant characteristics. The research team was familiar with these participants, as they were involved in our previous PWS behavioural studies. Four individuals were enrolled in previous clinical trials at sites in the United States. We used purposive sampling (Crossman,2020) to deliberately recruit individuals who we surmised would be cooperative, interested in the study, and had the verbal and cognitive skills to respond to the interview. Interviews were conducted in 2019–2020.

Prior to data collection, the study goals and procedures were explained to parents and individuals with PWS. Consistent with IRB policies, parents provided written, informed consent while those with PWS offered written and verbal assent. To protect anonymity, we used pseudonyms of participants.

### 2.2 | Procedures

Two clinicians with expertise in PWS administered a semi-structured interview to participants via Zoom. This on-line platform allowed

**TABLE 1** Demographic characteristics of 21 participants with PWS

| Characteristic | % or $M$ |
|---|---|
| Age | $M$ = 20.71; SD = 7.70; range = 14–38 years |
| Body mass index (BMI) | $M$ = 28.52; SD = 8.85; range = 19.0–43.6 |
| BMI classifications | Normal = 35%<br>Overweight = 25%<br>Obese = 40% |
| Gender | 43% M, 57% F |
| In school | 52% |
| Graduated | 48% |
| Living at home | 100% |
| Verbal IQ level | |
| Average | 20% |
| Low average | 35% |
| Borderline | 25% |
| Mild intellectual disability (ID) | 20% |
| Composite IQ level | |
| Average | 10% |
| Low average | 30% |
| Borderline | 40% |
| Mild ID | 5% |
| Moderate ID | 15% |
| PWS genetic subtypes | |
| Type I deletion | 19% |
| Type II deletion | 38% |
| mUPD | 33% |
| Unique deletion | 5% |
| Imprinting defect | 5% |

Abbreviations: mUPD, maternal uniparental disomy; PWS, Prader-Willi syndrome.

recorded, face-to-face interactions between participants and interviewers. All participants were interviewed at home. The interviews lasted between 30 and 60 min.

#### 2.2.1 | Semi-structured interviews

The three-part, semi-structured interview provided an organisational framework for data collection and leeway for interviewers to clarify or further probe responses. First, participants were asked how they think PWS affects them and their everyday life. Then, depending on their initial responses, interviewees were asked if weight, anxiety or outbursts were problematic for them. The third section probed participants' ideas about new medications for PWS, and how these might impact them. Finally, participants were invited to add anything else that they would like others to know about PWS, including drug companies.

#### 2.2.2 | Group versus individual interviews

Given differences in the schedules and availability of participants, 53% were individually interviewed, and 47% participated in group interviews with one to two others. As interviewers structured the group sessions in ways that did not foster extensive discussion among interviewees, we did not analyse group dynamics or processes as one does in traditional focus groups (Barbour, 2005).

#### 2.2.3 | Applied thematic analyses

Interviews were analysed using applied thematic analyses, a flexible methodology driven by the content of transcribed interviews (e.g., Braun & Clarke, 2006; Guest et al., 2012). Recorded interviews were transcribed verbatim, and entered into a secure, centralised database. The authors then read and re-read the transcripts, informally noting significant comments and possible codes or themes. Once familiar with the data, significant statements were extracted from the transcripts, including which section of the interview they were extracted from (Colaizzi, 1978). These 257 significant statements ranged in length from a few words to multi-sentence thoughts.

All phrases or sentences were numbered, allowing a clear audit trail for enhanced accuracy. A codebook reflecting specific content was developed, and after an initial round of coding, it was then revised and condensed. Revised codes were reviewed and discussed by the authors until coding consensus was achieved for the 257 significant statements. We then revisited the full transcripts to ensure that no significant statements were missed. Finally, themes and subthemes were established through an iterative process with the authors.

To enhance trustworthiness of findings, we established a clear audit trail, used a collaborative team approach, and also solicited a review of findings by outside experts. These experts included the Director and of Research and Director of Behavioural Studies at the Foundation for Prader-Willi Research (also the parents of young adults with PWS).

#### 2.2.4 | Cognitive functioning

As part of a previous longitudinal study, participants were administered the Kaufman Brief Intelligence Test-2 (KBIT-2; Kaufman & Kaufman, 2004) multiple times. While repeated IQ scores fluctuated between 0 and 8 points, corresponding classifications of cognitive functioning remained stable; IQ classifications were thus used in analyses (see Table 1).

#### 2.2.5 | Quantitative analyses

Frequency counts (presence or absence) of interview themes were calculated for the sample. Chi-square analyses determined if overall ability to respond to the interviews and the frequency of coded themes were associated with composite or verbal IQ classifications, gender, BMI or PWS genetic subtypes.

## 3 | RESULTS

### 3.1 | Overall response to interviews

Most participants (71%) were highly responsive and engaged with the interviews. Six individuals, however, had difficulty either understanding the questions or expressing their ideas. Interviewers used several scaffolding strategies with these participants (e.g., pausing, giving ample time to talk, slowing down and simplifying language, repetition). Despite these efforts, their responses remained sparse and lacked spontaneity. They were apt to repeat prompts from the interviewers or state, 'I don't know.' As such, these six individuals were not included in thematic analyses, leaving 15 respondents.

These six, less-responsive participants were all female, and differed in gender from their counterparts, $\chi^2$ (1, $N = 21$) = 6.30, $p < .01$. They did not, however, differ from others in PWS genetic subtypes, age, or cognitive functioning. Regarding IQ levels, 66% of non-responders and 60% of responders had Verbal IQs in the Borderline or Mild Intellectual Disability range. Composite IQs in the Borderline or Mild Intellectual Disability were also similar across non-responders versus responders (50% and 47%, respectively).

### 3.2 | Applied thematic analyses

Results are organised by: (1) the meaning and effects of PWS, which included three themes and five subthemes; and (2) opinions about what people with PWS need, which included two themes and five subthemes. Table 2 presents these themes and subthemes, as well as the percentages of participants who expressed them at any point during their interviews. Direct quotes from participants are italicised, along with their pseudonyms.

### 3.3 | Theme 1: Struggles with hunger and food seeking

All respondents indicated that, for them, PWS meant living with persistent hunger, and urges to food-seek and eat. As illustrated in Table 3, these young people offered poignant insights into how they experienced hunger as an omnipresent, distressful force that was enormously difficult to ignore; as Emily offered 'Food calls out to me, I can't think of anything else until I eat it'.

Participants used negative, emotionally charged language to describe their hunger and need for food (e.g., really hard, awful, upsetting), as well as the gravity of their condition (e.g., life-threatening, like cancer, food is a poison). Their comments also juxtaposed a longing to eat with the knowledge that one should not do so, for example, 'I got to eat something, but I can't' (Max).

**TABLE 2** Percentages of participants expressing themes and subthemes within two broad categories

| Broad category I: PWS means | Subthemes | % |
|---|---|---|
| Theme 1: Struggling with hunger, food and eating | | 100.0 |
| | 1a. Weight loss is hard | 47 |
| | 1b. Negative impacts on relationships, life choices | 61 |
| | 1c. Strategies to address | 61 |
| Theme 2: Struggling with anxiety, stress and outbursts | | 84 |
| | 2a. Schedule changes and other triggers | 67 |
| | 2b. School as a stressor | 67 |
| Theme 3: Distancing from PWS | | 61 |
| **Broad category II: PWS needs** | **Subthemes** | **%** |
| Theme 4: New medicines with diverse impacts | | 100 |
| | 3a. Hunger | 80 |
| | 3b. Anxiety, outbursts | 67 |
| | 3c. Improved relationships | 47 |
| | 3d. Meeting life goals | 61 |
| | 3e. A Cure | 47 |
| Theme 5: Advocacy and awareness | | 47 |

Abbreviation: PWS, Prader-Willi syndrome.

#### 3.3.1 | Subtheme 1A: Weight loss is hard

In contrast to hunger, fewer individuals (47%) commented that their weight was problematic. Although participants offered that maintaining or losing weight was hard, their remarks were less negatively charged than their insights about hunger, suggesting that weight pales in significance relative to chronic hunger. Many participants (40%) pointed out how their families helped them with their weight by ensuring food security at home.

#### 3.3.2 | Subtheme 1B: Negative impacts on life choices, relationships

Many interviewees (60%) described difficulties going out in the community. As Kaylee noted, 'You can't go where they have food around'. And, most communities 'have food around'. James astutely observed 'Fast food, it's everywhere. I am surrounded by food I shouldn't eat'. As summarised in Table 4, others described how their hunger and food availability thwarted their relationships with peers or family members, as well as their employment, living or educational goals.

#### 3.3.3 | Subtheme 1C: Strategies to manage hunger

Beyond familial measures to ensure food security at home, 61% of respondents offered far-ranging strategies that they find helpful in

**TABLE 3** Quotes representing theme 1 and subtheme 1a: Struggles with hunger and weight

| Theme 1: Struggles with hunger, food seeking | Subtheme 1A: Struggles with weight |
|---|---|
| *At home, if food's left out, it's just calling to me. It kind of feels like I can't think of anything else until I eat it*. Emily | *I gained 13 pounds from that camp in a week. Oh, gosh, it was difficult enough to lose the weight*. Jacob |
| *Then in the morning, food is like waking me up, that's the problem with me*. Kody | *Like why did I gain, when I get a lot of exercise and ate 1,000 calories? It should be less*. Gabe |
| *You know that you have it and it is life threatening, like cancer, and PWS you have it forever*. Thomas | *It's hard but I just lost two pounds so I'm happy about that*. Max |
| *I just feel hungry. I just feel bad and I can't really have anything*. Gabe | *Like my parents lock the pantry and stuff and that's for me*. Taylor |
| *I got to eat something, but I can't. I get hungry and start to be upset and get angry. It calms me down by eating*. Max | *Parents tell your kids about PWS at the right age. Then you can help them manage food*. Kyle |
| *Like the food around me, it's like a drug that kills me. The food is a poisonous drug that kills me*. Thomas | *Yes, yes, my parents help me by locking things up*. Blake |
| *Fast food, it's everywhere. I am surrounded by food I shouldn't eat. Am struggling very hard*. James | *Even if I try to eat a little bit, I still gain from eating something that is fatty*. Sophie |
| *It's hard to ignore being hungry, I want to eat more*. Anna | *I am good at my own diet, I do well*. Douglas |
| *It's really, really hard not to food seek*. Sophie | |

**TABLE 4** Quotes representing subthemes 1B and 1C: Negative impacts and management strategies

| Subtheme 1B: Negative impacts | Subtheme 1C: Management strategies |
|---|---|
| *PWS affects my family because when I'm full of temptation, you can't do anything, and it affects everybody.*<br>*It just sucks the whole time.* Kody<br>*Like at any kind of family function it's really hard cause my parents tell me that I can't go to social things with food.* Blake<br>*People don't go out to eat cuz they have the hunger issue.* Kody<br>*PWS affects like cooking and living on my own.* Sophie<br>*PWS affects the jobs that you get and how things go in the community, you can't go where they have food around.* Kaylee<br>*PWS affects me mostly by socializing. Because school might do some social after school thing and there is usually gonna be food around.* Gabe | *Sometimes I put notes on my hands. To remember things. So, tomorrow there is a girl's track meet and there's going to be cookies. So, the note would warn me don't go to it.* Gabe<br>*I need help with laundry cuz it's in the kitchen and nope, I don't go in the kitchen. So, have the laundry out of the kitchen!* Sophia<br>*I think it would be helpful to find ways to explain PWS to other kids. To let them know how to help you, so they could say, hey, you're not supposed to take that.* Gabe<br>*It's out of my control, but you can learn things to help like eating snacks in between meals. And I know when my mealtimes are, so I feel hungry then.* Kody<br>*When I want to eat more, I let my stomach settle for 5 minutes.* Anna<br>*There is one job for us and that's the cash register where people order the food and you do the credit cards. But you can hand them the food, and not actually take the food and eat it. Yeah, that's for them to eat. You didn't pay for it and you need to give it to the other person.* Kody |

**TABLE 5** Quotes representing theme 2 and subtheme 2a: Anxiety and triggers

| Theme 2: Anxiety, outbursts | Subtheme 2A: Schedule changes, other triggers |
|---|---|
| *I am getting nervous throughout the whole day.* Sophie<br>*Anxiety, it is just overwhelming.* Katlyn<br>*I know for me it's definitely the stress and a lot of anxiety.* Jacob<br>*Being worried affects me a lot.* Blake<br>*Anxious or worried is a lot for me. Like every single second of the day. Like I'm just worried about something until like it actually happens or gets resolved.* Emily<br>*Yes, ma'am, I do have a hard time controlling emotions.* Anna<br>*Controlling my emotions, yeah, that's kind of hard because sometimes I get mad easily.* Douglas<br>*Sometimes I just lose my temper on something or get emotional. It's like crazy the way I am talking or whatever.* Rose<br>*Like when I get stressed, I get angry at stuff. It's like when I get too out of control, I can do bad things. And then another bad feeling comes in like I could be out of my mind.* Max | *If somebody has a special thing that we don't know about, it's much more difficult for me because it throws a monkey wrench into my day.* Jacob<br>*Whenever there's a change, that gets me upset. When they just change it on me and not tell me. That gets me angry. When they tell me ahead of time, I'm fine.* Blake<br>*When they are late or things change, that's not a big deal if they tell me I don't get upset.* Max<br>*I got really mad at work. My task got changed. And it initially really shocked me. And then, that morning, the bus was late—really lateand I just couldn't handle it anymore, I guess. I was just full on tantrum mode.* Emily<br>*When my mentor is running late, I'm worried that she is not going to be able to take me out. Or if she, or if she's, sick, or if she's in traffic and I just don't know. And it's like waiting for hours for her to come and that gets me worried and anxious.* Blake<br>*When someone says no, I can't have this or I can't have that, I get upset.* Brody<br>*I get hungry and start to be upset and get angry.* Max<br>*My Mom starts things and then I get mad. My Mom does things to annoy me, even though she knows that it annoys me. Yeah, she does something and I get mad.* Douglas<br>*Sometimes I just take things the wrong way and then get angry or in a bad mood.* Rose |

managing their hunger or food seeking (see Table 4). Strategies included concrete steps (e.g., notes on hand, re-arranging the kitchen), enlisting help from peers, evoking social mores against stealing food, and ways of eating that they found helpful.

## 3.4 | Theme 2: Struggles with temper outbursts, stress and anxiety

Approximately 84% of participants indicated that for them PWS meant problems controlling their anxiety, anger and stress. As noted in Table 5, participants used a variety of terms to describe their emotional distress: stressed out, worried, anxious, angry, crazy, upset, mad, bad, and occasionally sad or unhappy. Individuals offered that they were anxious or worried 'a lot', that it was 'overwhelming' and occurred the 'whole day' or 'every second of the day'.

### 3.4.1 | Subtheme 2A: Schedule changes and other triggers of anxiety and outbursts

Some individuals noted that they just 'get mad easily' (Anna) and that temper outbursts were out of their control. Most participants (67%), however, attributed their worries or outbursts to a range of causes. As noted in Table 5, these triggers included feeling hungry or being

denied food, being provoked by a parent, faulty thinking, and unanticipated changes to their schedules or plans.

### 3.4.2 | Subtheme 2B: School as a stressor

Although schooling was not directly probed in the interviews, 67% of respondents described school as a stressful, anxiety provoking and predominantly negative experience. Notably, individuals who were no longer in school made over half of these negative comments, suggesting that schooling remained emotionally salient for them even as young adults.

While Katlyn offered that 'Everything about school is stressful', others identified specific aspects of school that they found especially problematic. As summarised in Table 6, these included: difficulties learning; homework and tests; being teased or treated poorly; the availability of food at school or in job training programs; and the use of food in lessons.

**TABLE 6** Quotes representing subtheme 2b: School as a stressor

| Difficulties learning | Tests, homework | Treated poorly | Food availability | Food in lessons, job training |
|---|---|---|---|---|
| *It is very hard to learn when you don't understand the material properly*. Sophie | *I get tests and then I get a bad grade and then I have to retake that test and then there's another test right behind that one and it keeps me up. The stress, the anxiety! Gabe* | *It could be like a teacher or kids being rude to me, sometimes they are mean*. Max | *It's really hard to stay with your diet, and truly pick wisely what you have for lunch*. Kody | *One of my teachers last year had a lot of labs and each one of them involved food*. Jacob |
| *When I'm at school, working on something I'm not sure how to do it, PWS affects the way I think or get things done*. Rose | *Like stress me out with homework!* Max | *Kids made fun of me and called me fat. That made me sad, made me down*. James | *We go past the lockers and I am like thinking as I go past them, I wonder if someone left food in there*. Gabe | *My teacher did a science experiment with Skittles, and she said she had to call my Mom. Like it's a life-threatening scenario!* Thomas |
| | | *I'd have to say, um, most guys don't accept me because of what I have*. Douglas | | *PWS affects me in my job training at school where they're preparing pizza*. Kody |
| | | | | *This job training program that I was doing, there was a lot of food around. I was sneaking food from the offices and asking for money. They weren't willing to work with me. I wouldn't recommend that program for any PWS student*. Jacob |

**TABLE 7** Quotes representing theme 3: Distancing from PWS

| Separates PWS from self | Denies, minimizes PWS | Better than others | Same as others |
|---|---|---|---|
| *It's like when I get too out of control, I can do bad things. Yeah, and it's not myself, it's the PWS*. Max | *I don't have it! I only have 3 pieces of it… three pieces is more like not having it at all!* Brody | *They really can't have hamburgers or hotdogs. They do it anyway. And what do you think happens? THEY GET FAT. Not me, I can eat anything!* Brody | *Losing weight? That's hard for everybody, even if they are normal!* Brody |
| *I won't be hungry, but my Prader-Willi will tell my brain that I'm still hungry*. Anna | *Some girls like fat guys!* Gabe | *Since my doctor told me that I'm one of the smartest ones, I watch what I eat. But it can be hard to be the smartest and not eat so much*. Max | *I want people to know we aren't really different; we are normal*. Gabe |
| | *I don't tell others about PWS, I keep it a secret*. James | *Some kids are tempted when they see something, they go and grab it and eat it, and that's not the case for me because I'm much better than that. PWS in mine works a little different*. Kody | |
| | *I have a hard time, and it's making me wish that I didn't have PWS because I don't like it at all. I shouldn't even have this chromosomal anomaly that I got*. Anna | | |

Abbreviation: PWS, Prader-Willi syndrome.

**TABLE 8** Quotes representing theme 4: What new medicines should do for PWS

| Hunger, weight | Anxiety | Family relations | Jobs, life goals |
|---|---|---|---|
| *I'd be able to have the food not locked up and not be hungry anymore*. Emily | *I would like medicine to be like not so anxious all the time. And not worry about things stressing me out as much*. Blake | *It would make me and my family happier, and we wouldn't have to be yelling or hurting each other*. Blake | *I would probably be able to have like a paying job*. Blake |
| *I could have like 10,000 calories!* Thomas | *I would like new medicine to control my temper. I would be happy*. Max | *Our home would be much less stressful. My parents would be happier. People would not get frustrated at me. We could have a nice time here at home*. Emily | *It would help people like us in the long run to be more independent. Like work at a job where there is food. I could go to school myself. I want to go to college*. Katlyn |
| *It is important to make just 1 medicine, not 7, that would help us lose weight. It would help PWS kids feel good about themselves*. James | *It would make my life better because you wouldn't have to worry about feeling anxious. I wouldn't be as unhappy as I am*. Anna | *My family would be much happier, they wouldn't punish you for all the stuff that you do*. Kody | *It would make it easier to live on your own*. Rose |
| *Everything wouldn't be locked. You could do stuff without battling yourself*. Gabe | *I'd want it to control the hunger and have some of the anxiety stuff in it too so it can help with the hunger and anxiety together*. Kody | *I wish it could make living in a house of people easier. I want to be more loving*. Anna | *I could work wherever I wanted to, even if it involved food. I'd be able to live on my own*. Emily |
| *It would be different because you are not thinking about food all of the time*. Kody | | | *I could do stressful things like school* Kaylee |
| *I could do things even when food is around*. Jacob | | | I *could live in an apartment and be a veterinary assistant*. Sophie |

| Medicines as a cure for PWS |
|---|
| *Cure the hunger and completely cure the urge to seek food*. Jacob |
| *The cure for us is a lot of things*. Sophie |
| *I want something to help with everything!* Thomas |
| *A new medicine for PWS should almost get rid of it*. Anna |
| *Help with the hunger! Foods! And anxiety! And emotions, too! All of them!* Emily |
| *They should make a lot of different types of medicines for all the different effects of PWS*. Gabe |
| *It would be a cure. Everyone could have the cure*. James |

Abbreviation: PWS, Prader-Willi syndrome.

**TABLE 9** Quotes representing theme 5: Awareness and advocacy

| Theme 5: Awareness and advocacy |
|---|
| *A new medicine needs to help all the PWS obesity in the world!* James |
| *I want the drug makers to allow everybody the opportunities to do the studies. A lot of them I can't do because I'm not in that age group*. Jacob |
| *I want them to know about PWS and spread it all over the news!* Thomas |
| *No one knows about it. There's even the butterfly syndrome where the skin cracks every time it moves. But PWS doesn't go up. If people were aware about it, they would start making new medicines*. Kaylee |
| *I want everyone to know that people with PWS can talk. They can. Like the drug companies were saying, my Mom was saying that people with PWS can't explain things. They can. They can say what they need and want and what's hard for them. Their parents don't need to do it. They can explain it*. Emily |

## 3.5 | Theme 3: Distancing from Prader-Willi syndrome

A theme emerged from 61% of respondents that reflected paths for how they may be incorporating PWS into their broader sense of self. Although varying in content, all of these paths represented efforts of participants to distance themselves from having PWS.

As shown in Table 7, some individuals denied having PWS, wished it away, kept it a secret, or minimised the effects of being overweight. Others split the syndrome off from who they really are, suggesting that PWS takes them over and is responsible for their outbursts or overeating. Still others distanced themselves by asserting that they were the same as other people or better than their counterparts with PWS.

## 3.6 | Theme 4: Medicines that 'cure', and lead to positive life outcomes

All participants readily endorsed the need for new medications to treat PWS (see Table 8). Most individuals, 80%, expressed the need for medicines that controlled their hunger and that could make it easier to lose weight. Still others described that 'Life would be different because you are not thinking about food all of the time' (Kody), or as Gabe expressed, 'You could do stuff without battling yourself'. Many participants, 67%, also voiced that new medicines should make them happier and less cranky, stressed out, anxious and worried.

Many respondents, 47%, described how reduced hunger or anxiety could positively impact their relationships. They offered that family conflicts would lessen, and family members would be happier and less stressed. Two individuals also voiced hopes that new medicines would help them 'Make more friends' (Max) or 'Be a lot more confident asking women out' (Douglas).

Similarly, 60% of participants described other positive, downstream effects of new medicines in meeting their life goals. These included living independently, having more options for employment, and being able to attend college or job training programs.

Interviewers did not use the word 'cure' in their probes, nor did they imply that new medicines would be curative. Even so, 47% of respondents framed new medications as a cure for PWS. While some individuals used the word 'cure', others implied a cure by stating that new medicine should help with all of the symptoms of PWS, and 'almost get rid of it' (Anna).

### 3.7 | Theme 5: Awareness, advocacy

Several interviewees (47%) expressed needs for greater awareness of PWS among the public and pharmaceutical companies. In doing so, they understood that their condition is rare, and that increased knowledge is an important step toward new drug development. As shown in Table 9, one young man advocated that people of all ages be included in future clinical trials. Another participant disagreed with clinical trials that use parents as informants and passionately stated that drug companies need to know that 'people with PWS can speak for themselves' (Emily).

### 3.8 | Quantitative analyses of themes

Chi-square analyses or Spearman's Rho correlations revealed no significant effects of age, PWS genetic subtype, BMI status or cognitive level for any themes or subthemes. A significant gender difference, however, emerged for Theme 3, with males approximately four times more likely than females (68% versus 17%) to distance themselves from PWS, $\chi^2$ (1, $N = 15$) = 3.62, $p < .05$.

## 4 | DISCUSSION

Participants offered novel insights into how they perceive and live with PWS. Although findings shed light on the lived experiences of those with PWS, they also have important implications for current management strategies and future clinical trials.

### 4.1 | Theme 1: Struggles with hunger and food seeking

Interviewees viewed the world around them through a window of hunger. They cast hunger and food seeking as omnipresent, and in negative, distressful terms that suggested that they were keenly aware that their syndrome was serious and life-threatening.

Participants also described the vicious cycle they experience of wanting food but knowing that they should not or cannot give into 'hunger urges. In this vein, it is impressive that many devised their own strategies for curbing food-seeking. Although speculative, doing so may foster feelings of self-efficacy and developmentally, young people with PWS may be more receptive to using to using peer-generated strategies than those offered by parents or dieticians.

Insights into the deleterious effects of chronic hunger in PWS can be gleaned from studies of hunger among people in general. People who are hungry for diverse reasons have difficulties sustaining attention on food-irrelevant tasks, thereby compromising their general problem-solving and planning abilities (Al-Shawaf, 2016). At the same time, hunger enhances memory of food stimuli (Montagrin et al., 2019), and the ability to solve food-acquisition problems (Al-Shawaf, 2016). Such findings are consistent with the ingenious food-seeking strategies used by many with PWS.

Although mild-to-moderate hunger is associated with impulsivity, irritability, aggression, distress and anxiety (Al-Shawaf, 2016; McLaughlin et al., 2012; Weinreb et al., 2002), it also impacts more basic emotional arousal and processing. Montagrin et al. (2019) found that hungry versus sated individuals were highly aroused by neutral stimuli, and thus had difficulties discriminating between emotionally intense versus neutral stimuli. Hungry people with PWS may similarly be aroused or overreactive to neutral stimuli, perhaps contributing to their anxiety, outbursts, or tendency to misinterpret benign social stimuli or interactions as ill-intended (Dykens et al., 2019).

At first glance, considerable overlap exists in the cognitive and psychological sequalae of hunger in the general population and symptoms of PWS. Yet it is challenging to disentangle symptoms of PWS and the extent to which they relate to hunger per se versus other genetic, cognitive, developmental, neural or environmental factors. As one participant, Kody, stated, 'It's hard to figure out if it is just the hunger urges or if the hunger and anger issues and anxiety all go together. Yeah, it's hard to figure that one out'.

### 4.2 | Theme 2: Struggles with anxiety, stress and outbursts

Almost all interviewees stated that PWS meant that they struggled to manage stress, anxiety and temper outbursts. Their statements revealed a high degree of emotional intensity that has yet to be adequately captured in the PWS literature, including studies of psychiatric disorders (Sinnema et al., 2011; Soni et al., 2007; e.g., anxious every second of the day, anxiety is overwhelming).

At the same time, interviewees generally had good insight into triggers for their anxiety or outbursts. While some blamed their outbursts on others, their syndrome, faulty thinking, or being denied food, several individuals noted that they became upset or anxious in the face of unexpected change. People with PWS typically exhibit high needs for predictability or sameness that are associated with deficits in executive functioning, especially in task switching (Chevalere et al., 2015; Woodcock et al., 2009). Task switching underpins cognitive flexibility and the ability to respond and adapt to changing situations. Deficits in these processes in people with PWS contribute to their need for predictability, repetitive questioning and temper outbursts (Woodcock et al., 2009, 2011).

Although two individuals offered that they did not get upset when they were informed of a schedule change, unpredictable events typically arise in the flow of everyday living. As such, other strategies are needed to de-escalate spiralling anxiety or outbursts in the face of

unpredictable change. Promising interventions include introducing low-stake changes to routines or schedules and playing novel computer games designed to enhance task-shifting abilities. Although studies are needed, programs aimed at increasing cognitive flexibility for individuals with ASD (e.g., Cannon et al., 2018) may also benefit people with PWS.

School emerged as a significant source of anxiety and stress for many participants; even those no longer in school were negative about their educational experiences. Accommodating to the needs of students with PWS is challenging for many school systems, especially given the rarity of this syndrome and the omnipresence of food in the school environment. Yet interviewees indirectly offered several ways that educators could think anew about students with PWS. It is important, for example, for educators to understand that the mere presence of food at school may distract students with PWS from staying attentive and focused on learning (e.g., 'I wonder if somebody left food in the lockers' Gabe). Educators could adjust their instruction to minimise food in lesson plans and identify job training programs outside of the food-service sector. Students with PWS should also be assessed for heightened anxiety or stress associated with learning goals and assignments (Roof, 2013).

### 4.3 | Theme 3: Distancing from Prader-Willi syndrome

All interviewees were well aware that they had PWS. Even so, 61% described ways that they distance themselves from their syndrome, suggesting that PWS holds a negative valence for them. Unexpectedly, males were four times more likely than females to engage in these distancing tactics. A gender difference was also reported by Cunningham and Glenn (2004) in their study on the self-perceptions of adults with Down syndrome. Specifically, males were more apt to express negative, avoidant reactions to their syndromic status, which the authors attributed to males having higher rates of negative social experiences. Although further work is needed, three participants in the current study, all males, described being treated poorly by others.

Participants' self-perceptions were both similar to and different from others with intellectual disabilities. Investigating the self-presentations of adults with intellectual disabilities, Finlay and Lyons (2000) found that respondents predominantly engaged in lateral or downward comparisons to others. Like some individuals in the current study, Finlay and Lyons' participants emphasised how they were the same as others or better than their peer group. Such social comparisons are seen as ways to avoid the stigma associated with intellectual disabilities and to facilitate a more positive self-representation (Beart et al., 2005). Other studies find that adults with intellectual disabilities are aware of their disability status but do not deem it important or central to who they are, instead emphasising their competencies and the activities, goals and relationships that are important to them (Björnsdóttir & Traustadóttir, 2010; Dorozeko et al., 2015; Kittelsaa, 2014).

In contrast to these studies, however, no participant with PWS used the word disability to describe themselves, nor did the transcripts include any mention of intellectual or developmental disabilities. Participants may have been unaware of these labels, aware of them but deemed them irrelevant, or the demand characteristics of the interview, with its focus on PWS, may have overshadowed any mention of disability. Nevertheless, findings raise the intriguing possibility that people with PWS identify primarily with their syndrome, and not necessarily with the clinical, research or social designations of PWS as an intellectual or neurodevelopmental disability.

Even so, and similar to people with other genetic conditions (Klitzman, 2009), many participants seemed to be wrestling with how, or if, to incorporate their syndrome into their broader sense of self. Developmentally, young people in general are prone to question 'Who am I?' Those with PWS or other syndromic diagnoses are also tasked with making sense of an assigned versus chosen aspect of their identity, and to question 'Am I my syndrome?' or 'Who am I in relation to this syndrome?' (Dykens, 2021). Unlike those with acquired disabilities, some scholars posit that people with congenital disabilities normalise their impairments because they are living with a condition they have always had (Beauchamp-Pryor, 2011; Shakespeare, 2006). Still, people develop a sense of self by making social comparisons and identifying how they are the same as or different from others (i.e., Finlay & Lyons, 2000; Stets & Burke, 2014). As Gabe noted, 'I want people to know we aren't really different, we're the same, but born with some difficulties'.

### 4.4 | Themes 4 and 5: Prader-Willi syndrome needs new mediations, clinical trials and advocacy

All interviewees wanted new medications that could curb their hunger, anxiety or outbursts and all wanted to enrol in future clinical trials. Notably, Tsai et al. (2018) reported that parents of individuals with PWS also prioritised hyperphagia and anxiety as key targets for future clinical trials. Themes 4 and 5 have several important implications for future trials.

Participants described many positive downstream effects of new medicines on their daily lives and aspirations. If hunger or anxiety were no longer problems, they envisioned improved relationships with family and peers, being able to work or live independently, attend college or job training programs, and 'to do things even when food is around' (Jacob). Indeed, many framed new medicines as a 'cure' for PWS.

These findings highlight the need to manage the expectations of clinical trial participants, especially during the informed consent and assent processes. Participants need to understand that medicines will not automatically translate into a 'cure', make them more independent or able to get along with others. Instead, trials or medications should be framed as opportunities for people with PWS to obtain the support and cognitive, adaptive and interpersonal skills they need in order to realise their living, work and relationships goals.

Interviews also have implications for patient versus proxy informants in clinical trials. Emily asserted people with PWS 'can speak for themselves', reflecting the desires of people with intellectual disabilities to self-advocate. Yet interview data sound a cautionary note for doing so in PWS trials.

Despite deliberately recruiting individuals who we believed had the capacity to respond to the interview, six young women struggled to verbalise meaningful responses, even with extensive scaffolding. Level of cognitive functioning did not differentiate responders from non-responders, nor did age or PWS genetic subtype. Instead, other factors likely impeded their performance, including inattention; limited insight or capacity to self-reflect; problems recalling or organising information or difficulties with the social demands of the interview (Emerson et al., 2013). It would thus be risky to assume that all participants with PWS can provide meaningful self-report clinical trial data, or to use cognitive functioning as a proxy for their ability to do so.

Finally, interview data highlight the importance of hyperphagia versus weight or the Body Mass Index as an endpoint in clinical trials. In both the frequency and tone of their remarks, participants emphasised that 'hunger urges' were omnipresent, regardless of their weight. Although maintaining a healthy weight was important, participants overwhelming noted that hunger is the overarching force they must reckon with all the time. As such, many clinical trials have used the informant based Hyperphagia Questionnaire as a primary endpoint (Dykens et al., 2007; Fehnel et al., 2015). More objective indices of hyperphagia also hold promise for future clinical trials, including eye-tracking measures of food versus other stimuli in visual exploration tasks (Key & Dykens, 2018), and visual event-related potentials of food stimuli (Key & Dykens, 2008).

Several study limitations deserve mention. First, findings should not be interpreted as representing all young people with PWS. As participants had previously been enrolled in PWS research, others without such experiences may voice different concerns or opinions. Second, the sample was limited to youth and young adults. Consistent with previous work, however, we have much to gain by studying the behaviour and self-perceptions of older adults with PWS (Dykens, 2004, 2013). Another limitation is that self-perceptions were only assessed at one time point. Yet one's self-perceptions are not stable or monolithic; they are fluid, differentiated and change in response to situational and interactional demands (e.g., Rapley et al., 1998). As constructing, revising and maintaining a sense of self is a life-long process, studies are needed on how the self-perceptions of individuals with PWS change with advancing age and development. Future clinical trials also offer a unique opportunity to explore how attenuated hyperphagia or anxiety impacts the lived experiences and self-perceptions of people with PWS.

Despite these limitations, this study is the first to give voice to people with PWS, and how they perceive their condition and the need for new pharmacological treatments. Participants conveyed how hard they work every day to manage the many challenges of their syndrome. At the same time, they also envision future goals and a better life for themselves and their families, reminding us that people with disabilities are much more than their limitations or chromosomal anomalies (Dykens, 2021; Zigler, 2001).

## ACKNOWLEDGEMENTS

The authors thank the Foundation for Prader-Willi Research (FPWR) for their enthusiastic support of this research, including Theresa Strong, PhD, Lauren Roth, PhD and the FPWR Clinical Trial Consortium. We are grateful to Robert Hodapp, PhD for his comments on an earlier draft of the manuscript. We also thank all study participants for sharing their time and important insights with us.

## DATA AVAILABILITY STATEMENT

The data that support the findings of this study are available from the corresponding author upon reasonable request.

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**How to cite this article:** Dykens, E. M., Roof, E., & Hunt-Hawkins, H. (2022). 'The cure for us is a lot of things': How young people with Prader-Willi syndrome view themselves and future clinical trials. *Journal of Applied Research in Intellectual Disabilities*, *35*(2), 460–470. https://doi.org/10.1111/jar.12950