




Biological, Behavioral, and Ethical Considerations of Prader-Willi Syndrome: A Primer for Behavior Analysts

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Abstract

Prader-Willi syndrome (PWS) is a rare genetic disorder typically characterized by hyperphagia, hypotonia, intellectual disabilities, insistence on routines, and obsession and compulsion related to food. Although current medical interventions primarily include growth hormones to address the biological symptoms of the individual, behavioral therapy is an alternative option for skill acquisition and decreasing problem behaviors. There is a growing need for applied behavior analysis (ABA) research on targeting problem behaviors and teaching requisite skills to individuals with this syndrome. This article reviews the current literature on PWS, highlights treatments and their limitations, suggests how ABA providers can provide ethical services, and proposes future research needs with this syndrome.

Keywords behavior analysis · clinical collaboration · Prader-Willi syndrome

Prader-Willi syndrome (PWS) is a rare genetic disorder occurring in roughly 1 in 22,000–25,000 births and an estimated 1 in 54,000–76,000 of the total population (Dudley et al., 2008; Khan et al., 2016). PWS presents with a characteristic phenotype and complex biological composition. There are consequential health issues and behaviors that arise with these common biological differences: hypotonia (low muscle tone), hyperphagia (insatiable appetite), growth hormone deficits, hypogonadism (small external genitalia), cryptorchidism (undescended testes) and mild to moderate intellectual disability (ID; Cassidy & Driscoll, 2009; Chen et al., 2007; Griffiths et al., 2014; McAllister & Whittington, 2011; Spindel, 2011). Other physical characteristics include short stature and prominent facial features (Cassidy & Driscoll, 2009; Griffiths et al., 2014; Spindel, 2011).

Additional commonly associated medical issues include inadequate sucking reflex during infancy, decreased arousal, and delays in reaching developmental milestones. These biological aspects are often seen in concert with skin picking/self-injurious behaviors, compulsive behavior, insistence on routines, food stealing, and overeating (Bennett et al., 2015; Chen

et al., 2007; Griffiths et al., 2014; Haig & Woodcock, 2017; Ho & Dimitropoulos, 2010; Spindel, 2011)—all of which are socially significant behaviors that may impede the individual's safety and quality of life. An average of 25% of individuals with PWS are diagnosed with autism spectrum disorder (ASD; Bennett et al., 2015; Veltman et al., 2004) with similar symptomatic overlap, such as ritualistic and routine-oriented behaviors (Bennett et al., 2015; Greaves et al., 2006; Veltman et al., 2004). With a variety of individual (i.e., biological, behavioral, environmental) and logistical (e.g., coordinating care across several support professionals from doctors to teachers and parents) considerations, this may pose challenges for a board certified behavior analyst (BCBA) working with an individual with PWS. Behavior analysts must consider how these challenges could impede their ability to help create successful environments dictated by the needs and wants of the individual.

Given the complexity of PWS, the purpose of this article is to address these biological and behavioral concerns, in particular, in relation to how applied behavior analysis (ABA) practitioners can take an active role within a team of service providers to create environments of success for individuals with PWS. Clinical and ethical considerations will be reviewed concurrently with practical suggestions for BCBA and service providers through reviewing current research. The article also highlights general limitations of the current behavioral research while providing suggestions for future areas of inquiry that both practitioners and researchers may consider.

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Biological Background and Medical Considerations

The diagnosis of PWS is dependent upon clinical observations and genetic testing (Cassidy & Driscoll, 2009; Ho & Dimitropoulos, 2010; Kundert, 2008) because there are several genetic subtypes: paternal microdeletion, maternal uniparental disomy (mUPD), and imprinting defects (Bennett et al., 2015; Cassidy & Driscoll, 2009; Griffiths et al., 2014; Irizarry et al., 2016)—each with differing effects on an individual's overall developmental repertoire. There are noted phenotypic discrepancies among the three subtypes that affect an individual's learning and behaviors. Those with the paternal deletion subtype are more likely to engage in skin picking and less likely to manifest with psychiatric concerns in comparison with mUPD (Cassidy & Driscoll, 2009; Kundert, 2008; McAllister & Whittington, 2011). In this subtype, a comorbid diagnosis of ASD is more common in comparison to paternal microdeletion and genetic imprinting defects (Ho & Dimitropoulos, 2010; Whittington & Holland, 2010).

Further, individuals with PWS are likely to experience hypothalamic abnormalities (Cassidy & Driscoll, 2009; Griffiths et al., 2014; Irizarry et al., 2016; Khan et al., 2016; Kundert, 2008). These abnormalities in the hypothalamus are related to a range of medical and behavioral effects, such as energy expenditure (e.g., hyperactivity), hunger and thirst regulation (e.g., increased hunger and food-seeking behaviors), muscle tone (e.g., making daily living skills such as self-care difficult), sleep disturbances, pain tolerance, and temperature insensitivity (Griffiths et al., 2014; Irizarry et al., 2016; Khan et al., 2016; Kundert, 2008; Whittington & Holland, 2010).

Perhaps the most socially significant consequences from hypothalamic irregularity are lack of satiety with food (i.e., hyperphagia), food seeking, and a preoccupation with food leading to potential overeating and food stealing, which may present personal (e.g., overeating) and social (e.g., isolation) consequences depending on the environmental expectations within the individual's life (Cassidy & Driscoll, 2009; Griffiths et al., 2014). Hyperphagia has been linked directly to obesity (Holsen et al., 2006; Whittington & Holland, 2010), and can lead to lifelong struggles when in combination with hypotonia (Bennett et al., 2015; Khan et al., 2016). Increased exercise to counteract high caloric intake may be difficult or even painful given the low muscle tone and, as a result, fatigue associated with hypotonia (McAllister & Whittington, 2011). In addition, around 20%–25% of adults with PWS have Type II diabetes, therefore, concerns related to food intake potentially pose a large health risk to the individual (Cassidy & Driscoll, 2009; Khan et al., 2016; Whittington & Holland, 2010).

Another medical consideration experienced by individuals with PWS is decreased arousal, defined as a lack of energy (such as fatigue or decrease in sexual activity), primarily due

to hypotonia (Gross-Tsur et al., 2011). Low muscle tone is associated with achieving developmental milestones later in life, making hypotonia a socially significant aspect of PWS (Cassidy & Driscoll, 2009; Whittington & Holland, 2010). In particular, the effects of hypotonia can be challenging for those who engage in intimate interactions with consenting partners.

Physicians often address hypotonia through growth hormone treatments (Dykens et al., 2017). In a study by Dykens et al. (2017), growth hormone therapy was found to increase verbal and daily living skills, and more so when treatment began before the age of 12 months. With the use of these hormones, individuals with PWS may experience increased levels of energy and muscle tone, allowing them to participate in more exercise routines, which may contribute to reduction in potential weight concerns. It should be noted that the hormones alone cannot increase an individual's preference for exercise and a behavioral intervention may be more effective to address obesity than medical intervention (Dudley et al., 2008). When working on a team serving an individual with PWS, service providers should work closely with the medical professionals on that individual's team and note any behavioral changes or adjust protocols when hormone treatment begins. It is of note that BCBA's should not suggest medical treatment because this is not within the scope of practice (or competence) for most BCBA's, save for individuals who may be medical professionals in addition to having a BCBA credential (even in this scenario, they would be operating under a medical license and not as a BCBA). What behavioral practitioners may do, however, is work in concert with the medical professionals on their client's team in order to (1) note behavioral or developmental changes with any new medication regimens, (2) address potential areas of increased health need (e.g., exercise regimens) as detailed by the individual's medical professionals within the limits of what is comfortable for the individual themselves, and (3) provide medical or nutritional adherence programming through behavioral skills training (BST) for not only the individual with PWS but their family and other service providers as well.

Common Behavioral Concerns and Relevant Approaches

Hyperphagia and Food-Stealing

In clients diagnosed with PWS, hyperphagia (e.g., eating more than socially appropriate) is a common target behavioral practitioners address due to its significant impact on the client's health and social relationships (Ho & Dimitropoulos, 2010). Common protocols for targeting eating-related behaviors are locking up the food and 24/7 food monitoring (Griffiths et al., 2014; Ho & Dimitropoulos, 2010). At first, the use of

restrictive procedures is effective at protecting the safety of the client and reducing the amount of food consumed. However, behavioral practitioners must consider the long-term sustainability of these restrictive methods, particularly in relation to generalization and maintenance. Restricting food from an individual brings up a variety of ethical challenges and, if done incorrectly and without monitoring, can be extremely harmful for an individual and should only be considered with consistent oversight by behavioral (e.g., BCBA, social workers) and medical (e.g., MD, nutritionist) practitioners to ensure healthy weights are maintained and the client's autonomy is minimally affected. If this tactic is used as an initial procedure, a plan should be devised for when to fade this restrictive approach and other, more sustainable, approaches can be used long term.

Although restricting access to food to prevent overeating may seem effective in some environments, it does not take into consideration food-stealing behaviors in more communal environments where food is openly available (e.g., cafeterias, larger households). There are a variety of alternatives to locking up food that include less intrusive or preventative methods.

Page et al. (1983) who used a differential reinforcement of other behavior (DRO) procedure to reduce the amount of food stealing behaviors in clients with PWS. In this study, reinforcement occurred on an interval schedule and increased using progressive intervals that resulted in a significant decrease in food stealing behaviors for all three participants in their respective therapeutic settings. However, the reduction on food stealing behavior did not generalize to other settings (Page et al., 1983) and future research should consider expanding treatment in different settings across different people (i.e., behavior analysts, caregivers, teachers). This study suggests that food removal may be useful in the short term to decrease food-stealing behaviors but the sustainability of the intervention is in question.

Maglieri et al. (2000) successfully treated covert food-stealing in a 14-year-old individual with PWS. The intervention established stimulus control by using labels on food containers and punishment in the form of verbal reprimands. Treatment took place in a hospital setting using a kitchen-like therapy room with a refrigerator, and leisure materials were available. A one-way mirror allowed the therapists to leave the participant alone with the food while continually monitoring her. During baseline, cookies and pretzels were left in the room with her while the therapist left. The participant was told not to eat the food, but there were no consequences regardless of her behavior. Next, the authors implemented a within-session reprimands condition (Maglieri et al., 2000). Contingent on the participant stealing food, the therapist came in, verbally prompted her to drop the food and reprimanded her, such as, “that’s not your food, stop eating it” (Maglieri et al., 2000, p. 616). In addition, a scale was used

to weigh the food in front of the participant; if food had been taken, she was immediately verbally reprimanded. The authors then implemented a stimulus discrimination component, using orange stickers to label separate containers with prohibited food. The participant was shown and told she could eat food from the nonlabeled containers (those without stickers). Although covert food-stealing behavior decreased for this individual following the intervention in this study, a variety of ethical considerations, and potential violations arise.

Punishment procedures, such as verbal reprimand, widely range in both procedure type and effectiveness. In a literature analysis of 22 empirical articles, Pokorski and Barton (2020) discovered that most punishment procedures implemented were able to decrease overall target, aberrant behaviors. The additional, and perhaps more pertinent finding was that many of these studies did not adhere to ethical guidelines suggesting that although these approaches may have “worked,” they may have unintentionally put the research participants in prolonged aversive situations or states. When applying to verbal reprimands and other punishment-based procedures to decrease food stealing, the ends may not justify the means. It is important to respect the individuals' autonomy as well as respect for being—if a verbal reprimand, or adjacent approach, is used, it may be useful for initial decrease but should be faded to less aversive procedures involving reinforcement-based approaches.

In one such intervention, Lambert et al. (2019) utilized a token economy to decrease food stealing in a 7-year-old girl with PWS providing a trial-based functional analysis that increased and decreased in latency throughout the study to determine the effectiveness of the treatment. Three major components of the intervention were response blocking (of attempts to steal food; always redirected manually to exchange a token), differential reinforcement of other behavior (DRO; no food stealing), and differential reinforcement of incompatible behavior (DRI; exchanging tokens; Lambert et al., 2019). The authors used a trial-based format throughout with the general procedure including removal of food with onset food-stealing establishing operations (EO) and ending after this behavior occurred or after a fixed period of time without the presence of food-stealing (Lambert et al., 2019). A 36-s time delay was meant to mimic the average duration of finishing her meals with that of the rest of her family. The authors completed token-board conditioning after 27 blocks of trials and completed treatment within 46 trials (Lambert et al., 2019). By the end of treatment, the participant waited for each token to earn and ask for food rather than engaging in food stealing. The treatment also successfully generalized with other individuals and in other settings (i.e., at home during family meals) suggesting that a procedure without the use of punishment can be utilized in order to decrease potential food-stealing behaviors. For the behavior analyst in practice, this could be utilized through staff implementation, as well as

training service and support members in the lives of individuals with PWS. Future research could focus on the training of these professionals as well as family members to create environments of success for more sustainable approaches to food-stealing behaviors beyond those based in punishment.

Self-Injurious Behavior(s)

Another significant behavior frequently observed in individuals with PWS is self-injurious behavior (SIB), in particular in the form of skin picking. In a 2014 study by Hall et al., experimental functional analyses were conducted for 13 individuals that engaged in daily skin picking that damaged tissue and included five conditions: ignore, alone, attention, play, and demand. The researchers included the ignore condition to determine whether or not an adult being present would evoke the skin-picking behavior (Hall et al., 2014). The skin picking primarily occurred during the alone and ignore conditions, indicating that skin picking was maintained by automatic reinforcement. When an individual engages in SIB that directly affects their health and well-being, behavioral practitioners should consult with the individual's care team (e.g., medical doctors, educational staff) by (1) documenting the severity in which skin-picking has occurred, (2) reviewing the potential behavioral programming to be implemented with relevant goals, and (3) training steps for the generalization of these skills to the team to best support the individual.

In addition, in a review by Lang et al. (2010) behavioral interventions for skin picking among other individuals with disabilities were analyzed for overall effectiveness and approach. Their review corroborates the hypothesized function of automatic reinforcement in skin picking with some interventions including differential reinforcement, escape extinction, competing activities, and fading of the protocols as the behaviors decrease. They also discuss punishment procedures such as overcorrection, response blocking, and time-out—but the use of reinforcement procedures were just as effective suggesting that punishment may not have to be used to reduce this behavior and reinforcement strategies can help to promote more inclusive, less aversive environments. A thorough review of Lang et al. (2010) regarding various behavioral treatments of chronic skin picking is highly recommended for practitioners so that a bevy of options can be pulled from with relevant assessment and resource analysis.

There currently are few research studies that demonstrate effectiveness of targeted behavioral intervention for individuals with PWS. Table 1 represents a comprehensive summary of the research currently available for review and extension in both clinical and research settings. Future clinical and research investigators alike should expand on the current approaches listed above as well as other behaviors that are not included in this prior research such as socialization goals, academic readiness, daily living skills, vocational activities, consent and assent approaches, and self-advocacy skills.

Ethical Considerations and Interdisciplinary Collaboration

When working with individuals with PWS, ethical dilemmas may arise even with reputable, empirically based treatments. These ethical dilemmas occur both within behavior analysis and with the use of medical interventions. For example, although early use of growth hormones has overall positive effects on an individual's quality of life and behavior(s), the overall lack of access due to cost, geographical availability, or a lack of local service providers adept at this approach makes obtaining the hormones difficult for all individuals (Gondoni et al., 2008; Prader-Willi Syndrome Association, 2020). Not all individuals with PWS have hormone therapy as part of their overall treatments they engage with. However, if the behavioral practitioners does have an individual with PWS who is receiving this type of therapy, they need to collaborate with the treating physicians and work closely to review any potential differences in development over time or if there are any interruptions in receiving this therapy as it may affect their overall effectiveness of behavioral programming—if someone had a medical regime change, this may have effects on their behavior and as such, this needs to be noted and shared with the care team. In addition, due to the significant biological components associated with maladaptive eating and self-injurious behaviors, practitioners must be especially informed and careful when making treatment recommendations.

Although there is currently limited research on effective treatment packages focusing on behavior-analytic interventions, there are a multitude of potential avenues for behavior analysts to participate in PWS behavioral research. Across areas of treatment and research, certified behavior analysts are required to follow the ethical rules and regulations created by the Behavior Analyst Certification Board (BACB, 2014). Table 2 lists BACB Ethical Codes for Behavior Analysts Relating to PWS Treatment for a visual guide of a few relevant codes to consider when providing treatment to individuals with PWS.

Multidisciplinary approaches increase the individual's likelihood of success, such as working alongside medical and developmental experts (Brodhead, 2015; Ho & Dimitropoulos, 2010; Kundert, 2008). Furthermore, beginning growth hormone treatment early as an antecedent strategy can increase physical mobility and prevent significant health issues later in life (Cassidy & Driscoll, 2009; Ho & Dimitropoulos, 2010). In clinical practice, behavior analysts should collaborate with the individual's medical team and note in the data when hormone therapy interventions begin, end, or change in order to properly address potential biological effects with behavioral manifestations.

When creating behavioral programming, behavior analysts should take into consideration development of the individual and working alongside disciplines such as occupational

Table 1 Summary of Areas of Concern and Potential Treatments for Individuals with PWS

Area of Concern	Associated Behaviors	Behavioral Approaches	Citations
Obesity: an excess of body fat mass; often measured using BMI	<ul style="list-style-type: none"> • Reduced physical activity • Low energy expenditure • Overeating 	<ul style="list-style-type: none"> • Restricting food access; see treatments below for hyperphagia • Physical exercise programs (studies are limited) • Dietary planning 	<ul style="list-style-type: none"> • Bellicha et al. (2019) • Dudley et al. (2008) • Dykens et al. (2007) • Griffiths et al. (2014) • Hoffman et al. (1992) • Holland et al. (1995)
Intellectual disability: limited/below average intellect, onset before age 22; generally, an IQ score of 75 or below	<ul style="list-style-type: none"> • Insistence on routines • Compulsive behaviors • Fluctuating moods • Academic difficulties and learning disabilities 	<ul style="list-style-type: none"> • Behavioral interventions should be tailored to each individual's level of functioning and hypothesized functions of behavior 	<ul style="list-style-type: none"> • Ho & Dimitropoulos (2010) • Greaves et al. (2006) • Griffiths et al. (2014) • Lambert et al. (2019)
Hypotonia: low muscle tone	<ul style="list-style-type: none"> • Decreased arousal • Poor suck during infancy • Low levels of physical exercise • Poor reflexes 	<ul style="list-style-type: none"> • Most often treated with growth hormone therapy rather than behavioral intervention • Physical exercise programs 	<ul style="list-style-type: none"> • Cassidy & Driscoll (2009) • Chen et al. (2007) • Holland et al. (1995) • Kundert (2008)
Hyperphagia: persistent hunger, insatiable appetite, decreased satiation-related to hypothalamic abnormality	<ul style="list-style-type: none"> • Overeating • Food stealing • Pica • Hoarding food • Foraging for food • Excessive food interest 	<ul style="list-style-type: none"> • Restricting food access • Differential reinforcement of permitted and prohibited foods • Token boards and schedule thinning • Punishment procedures such as verbal reprimands 	<ul style="list-style-type: none"> • Chen et al. (2007) • Dimitropoulos et al. (2000) • Ho & Dimitropoulos (2010) • Lambert et al. (2019) • Lang et al. (2010) • Maglieri et al. (2000) • Page et al. (1983)

Note. The information above provides general suggestions and summaries when developing treatment services. This is by no means an all-inclusive resource; these shortened explanations are for brevity. Further review of these articles will detail how to ethically and morally apply treatment packages.

therapy (OT). Failure to generalize and maintain behavior change may lead to difficulties for adults with PWS when transitioning to a more independent lifestyle (Crinò et al., 2016). As stated by Ho and Dimitropoulos (2010), adults with PWS in their (chronological) 20s have the highest rate of

maladaptive behaviors and social problems—attributed to difficulties making the transition into a more independent living situation without constant supervision. Collaboration with other professionals may help to alleviate some of these concerns. In Crinò et al. (2016), it is argued that when young

Table 2 BACB Ethical Codes for Behavior Analysts Relating to BACB Ethical Codes (BACB 2022) (2020)

BACB Ethical Code (2022)	Considerations
1.04 Practicing within a defined role	<ul style="list-style-type: none"> Identify how a behavior analytic approach might support the individual and whether or not behavioral services would be necessary given the individual's other services Clearly communicate with the individual's team regarding these potential supports Find the appropriate supervision if one does not currently have this Find the appropriate training followed by ongoing supervision Refer to appropriate professional if the above are not realistic with current time and resources
1.05 Practicing within scope of competence	
2.01 Providing effective treatments	<ul style="list-style-type: none"> Assess the individual's wants and needs for behavioral change Consult with individual's support structure (parents, teachers, service providers, etc.) on desired, socially significant goals Create programming reconciling the above with the medical and behavioral challenges exhibited by the individual
2.10 Collaborating with colleagues &	<ul style="list-style-type: none"> Consult with the individual's current medical team and note any changes in treatment in data to further analyze potential behavioral effects (e.g. What happens after hormone therapy begins? How does this change your treatment moving forward?)
2.12 Considering medical needs &	<ul style="list-style-type: none"> Consult with the individual's education and therapeutic teams to ensure consistency and compatibility across services and goals
2.15 Minimizing Risk of Behavior-Change Interventions &	<ul style="list-style-type: none"> Review treatment effectiveness, compatibility with other services goals, and individual consent of the protocols frequently throughout the therapeutic relationship
2.19 Addressing conditions interfering with service deliveries	
3.12 Advocating for Appropriate Services (1.04, 1.05, 2.01, 2.08) &	<ul style="list-style-type: none"> Review any discrepancies between goals of other service providers and behavior analytic goal to reconcile to the individual's benefit Given that PWS is a complex syndrome, if there is a place for behavior analytic services to create new goals or support other's goal, do so respectfully and ensure that suggestions are in the best interest of the individual and not only the support structure around them
6.03 Research in Service Delivery (see 1.02, 1.04, 2.01, 3.01)	

Note. The above table details pertinent BACB ethical guidelines and considerations specifically geared towards the complexities of PWS. It is by no means a complete list but only a starting point for a behavior analyst to consider.

Table 3 PWS Organizations and Supports Offered

Organization Name	Supports Offered	Source/Site Link
Prader-Willi Syndrome Association (PWSA)	<ul style="list-style-type: none"> • Access to free research, events and educational resources on PWS • Research on areas of hyperphagia, intellectual disability, and hypotonia • Provides family support through care packages, mental health support, and family testimonials 	https://www.pwsausa.org/
The Foundation for Prader-Willi Research	<ul style="list-style-type: none"> • Promotes continual research on treatments and interventions for those with Prader-Willi • Research on evidence-based treatments for clinicals 	https://www.fpwtr.org/about-prader-willi-syndrome#definition
International Prader-Willi Syndrome Organization (IPWSO)	<ul style="list-style-type: none"> • Connect those with Prader-Willi, their families, and professionals from across the globe • Worldwide research on current treatments and services available for those with Prader-Willi 	https://ipwso.org/membership/country-members/
Prader-Willi Syndrome Families United	<ul style="list-style-type: none"> • Not for Profit organization that connects families and those with Prader-Willis together • Fundraiser Events to support research on Prader-Willi 	http://pwsfamiliesunited.org/about-us/

Note. The above table reviews organizations that are important in the support of individuals with PWS, their families, and their care teams. “Supports Offered” are potential, but not all-inclusive ways that they be useful for behavioral practitioners can utilize these resources.

adults with PWS are transitioning between settings, multiple disciplines working together promotes a smoother transition.

Incorporating a dietician is of particular importance for those individuals with maladaptive eating behaviors. These professionals can provide recommendations for food intake, meal preparation and planning. Dietary planning could coincide with an exercise-focused behavioral intervention. A study by Hoffman et al. (1992) described weight management and meal preparation in group homes for those diagnosed with PWS, emphasizing the importance of including choice for individuals when dieting. Behavior analysts are well-equipped to provide choice-making programs to coincide with dietary decisions. Other disciplines to consider are physical therapy and speech-language pathology to assist with physical fitness and communication, respectively.

Conclusion

Although there have been several suggestions for future research and clinical paradigms throughout this article, there are others that are worth noting. Research on PWS continues to expand in areas such as physical exercise (Bellicha et al., 2019), the individual’s perception of emotions and social cues (Dykens et al., 2019), routine insistence and change resistance (Haig & Woodcock, 2017), rate of feeding (Rigamonti et al., 2014), and providing behavioral support and parent training via telehealth (Zyga et al., 2018). Limitations of the existing literature include narrow participant demographics and insufficient generalization within the population of PWS. In particular, the current research does not represent a diverse sample and does not adequately include adult populations (Bellicha et al., 2019; Bennett et al., 2015; Gondoni et al., 2008; Ho & Dimitropoulos, 2010; Lowe-Greenlee, 2019; Rigamonti et al., 2014; Spindelov, 2011). Longitudinal studies are infrequent (Bennett et al., 2015; Gondoni et al., 2008), which is considered a limitation for several reasons. From a behavior analytic perspective, generalization and maintenance are integral aspects of scientific studies, which in turn potentially lead to more effective, person-centered treatments.

There are a variety of areas in which further research is warranted given the findings of this article. From a medical standpoint, research on comorbidity treatments within PWS could benefit those individuals with multiple diagnoses (Bennett et al., 2015; Whittington & Holland, 2010). In addition, the long-term effects of growth hormone therapy are increasingly relevant as its use becomes more widespread. In addition to medical intervention, behavioral treatments with technological guidelines to manage hyperphagia could lower obesity-related deaths and increase quality of life (Bennett et al., 2015; Khan et al., 2016). Along with eating behaviors, the paucity of research around skin picking as it relates to individuals with PWS needs to be addressed. Increasing flexibility, utilizing schedules, and promoting socialization are all

well-researched topics within the field of behavior analysis and future research would do well to apply these same concepts to supporting individuals with PWS.

Much of the previous research on PWS has solely focused on the general effects of medical advances, and less on the interaction between those interventions with behavioral services. The combination of genotypical and phenotypical characteristics of this population uniquely affect behaviors, and therefore it is not sufficient to simply replicate previous interventions for similar behaviors but rather to expand on long lines of research that have targeted similar behaviors with other individuals with similar behavioral repertoires.

There are several contingencies to include while analyzing and forming treatment plans for those diagnosed with PWS, most notably the medical components that affect behavior, this article hopes to serve as a starting point—not an all-inclusive analysis—for behavior analysts to begin collaborating with a client’s care team should they work with individuals with PWS. Table 3 also provides, as of the writing of this manuscript, organizations the behavioral practitioners can connect with in order to best support their clients, including what types of supports may be offered. Many of these organizations are easily accessible, with some connecting you to providers that specialize in PWS in the area in which one would practice. This includes care coordination services, suggestions for collaboration, genetic consulting, and research lines currently running. Through questioning and expanding on current research while continuing to provide supervised and collaborative work with an individual’s entire care team, behavior analysts can develop treatments that have the potential to increase the quality of life as defined by individuals with PWS.

Availability of Data and Material All data and literature was freely available via EBSCO host and Google Scholar.

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Code Availability Not applicable.

Declarations

Conflict of Interest The authors declare no conflict of interest.

References

- Behavior Analyst Certification Board. (2014). *Professional and ethical compliance code for behavior analysts*.
 Behavior Analyst Certification Board. (2020). *Ethics code for behavior analysts*. Littleton, CO: Author.

- Bellicha, A., Coupaye, M., Léonore, H., Speter, F., Oppert, J.-M., & Poitou, C. (2019). Increasing physical activity in adult women with Prader-Willi syndrome: a transferability study. *Journal of Applied Research in Intellectual Disabilities*, 33(2), 258–267. <https://doi.org/10.1111/jar.12669>.
- Bennett, J. A., Germani, T., Haqq, A. M., & Zwaigenbaum, L. (2015). Autism spectrum disorder in Prader-Willi syndrome: a systematic review. *American Journal of Medical Genetics, Part A*, 167a(12), 2936–2944. <https://doi.org/10.1002/ajmg.a.37286>.
- Brodhead, M. T. (2015). Maintaining professional relationships in an interdisciplinary setting: Strategies for navigating nonbehavioral treatment recommendations for individuals with autism. *Behavior Analysis in Practice*, 8(1), 70–78.
- Cassidy, S. B., & Driscoll, D. J. (2009). Prader-Willi syndrome. *European Journal of Human Genetics*, 17(1), 3–13. <https://doi.org/10.1038/ejhg.2008.165>.
- Chen, C., Visootsak, J., Dills, S., & Graham, J. (2007). Prader-Willi syndrome: an update and review for the primary pediatrician. *Clinical Pediatrics*, 46(7), 580–591.
- Crinò, A., Fintini, D., Bocchini, S., Carducci, C., & Grugni, G. (2016). Prader-Willi syndrome: Clinical problems in transition from pediatric to adult care. *Research & Reports in Endocrine Disorders*, 6, 49–57. <https://doi.org/10.2147/RRED.S56401>.
- Dimitropoulos, A., Feurer, I. D., Roof, E., Stone, W., Butler, M. G., Sutcliffe, J., & Thompson, T. (2000). Appetitive behavior, compulsivity, and neurochemistry in Prader-Willi syndrome. *Mental Retardation 7 Developmental Disabilities Research Reviews*, 6(2), 125–130. [https://doi.org/10.1002/1098-2779\(2000\)6:2](https://doi.org/10.1002/1098-2779(2000)6:2).
- Dudley, O., McManus, B., Vogels, A., Whittington, J., & Muscatelli, F. (2008). Cross-cultural comparisons of obesity and growth in Prader-Willi syndrome. *Journal of Intellectual Disability Research*, 52(5), 426–436. <https://doi.org/10.1111/j.1365-2788.2008.01044.x>.
- Dykens, E. M., Maxwell, M. A., Pantino, E., Kossler, R., & Roof, E. (2007). Assessment of hyperphagia in Prader-Willi syndrome. *Obesity*, 15(7), 1816–1826. <https://doi.org/10.1038/oby.2007.216>.
- Dykens, E. M., Roof, E., & Hunt-Hawkins, H. (2017). Cognitive and adaptive advantages of growth hormone treatment in children with Prader-Willi syndrome. *Journal of Child Psychology & Psychiatry*, 58(1), 64–74. <https://doi.org/10.1111/jcpp.12601>.
- Dykens, E. M., Roof, E., Hunt-Hawkins, H., Daniell, C., & Jurgensmeyer, S. (2019). Profiles and trajectories of impaired social cognition in people with Prader-Willi syndrome. *PLoS One*, 14(10), 1–19. <https://doi.org/10.1371/journal.pone.0223162>.
- Gondoni, L. A., Vismara, L., Marzullo, P., Vettor, R., Liuzzi, A., & Grugni, G. (2008). Growth hormone therapy improves exercise capacity in adult patients with Prader-Willi syndrome. *Journal of Endocrinological Investigation*, 31(9), 765–772. <https://doi.org/10.1007/BF03349255>.
- Greaves, N., Prince, E., Evans, D. W., & Charman, T. (2006). Repetitive and ritualistic behaviour in children with Prader-Willi syndrome and children with autism. *Journal of Intellectual Disability Research*, 50(2), 92–100. <https://doi.org/10.1111/j.1365-2788.2005.00726.x>.
- Griffiths, D., Condillac, R. A., & Legree, M. (2014). *Genetic syndromes and applied behaviour analysis: A handbook for ABA practitioners* (pp. 225–256). Jessica Kinsley.
- Gross-Tsur, V., Eldar-Geva, T., Benarroch, F., Rubinstein, O., & Hirsch, H. J. (2011). Body image and sexual interests in adolescents and young adults with Prader-Willi syndrome. *Journal of Pediatric Endocrinology & Metabolism*, 24(7–8), 469–475.
- Haig, E. L., & Woodcock, K. A. (2017). Rigidity in routines and the development of resistance to change in individuals with Prader-Willi syndrome. *Journal of Intellectual Disability Research*, 61(5), 488–500. <https://doi.org/10.1111/jir.12368>.

- Hall, S. S., Hustyi, K. M., Chui, C., & Hammond, J. L. (2014). Experimental functional analysis of severe skin-picking behavior in Prader-Willi syndrome. *Research in Developmental Disabilities*, 35(10), 2284–2292. <https://doi.org/10.1016/j.ridd.2014.05.025>.
- Ho, A. Y., & Dimitropoulos, A. (2010). Clinical management of behavioral characteristics of Prader-Willi syndrome. *Neuropsychiatric Disease & Treatment*, 6, 107–118.
- Hoffman, C. J., Aultman, D., & Pipes, P. (1992). A nutrition survey of and recommendations for individuals with Prader-Willi syndrome who live in group homes. *Journal of the American Dietetic Association*, 92(7), 823–833.
- Holland, A. J., Treasure, J., Coskeran, P., & Dallow, J. (1995). Characteristics of the eating disorder in Prader-Willi syndrome: Implications for treatment. *Journal of Intellectual Disability Research*, 39(5), 373–381. <https://doi.org/10.1111/j.1365-2788.1995.tb00541.x>.
- Holsen, L. M., Zarcone, J. R., Brooks, W. M., Butler, M. G., Thompson, T. I., Ahluwalia, J. S., Nollen, N. L., & Savage, C. R. (2006). Neural mechanisms underlying hyperphagia in Prader-Willi syndrome. *Obesity*, 14(6), 1028–1037. <https://doi.org/10.1038/oby.2006.118>.
- Irizarry, K. A., Miller, M., Freemark, M., & Haqq, A. M. (2016). Prader-Willi syndrome: Genetics, metabolomics, hormonal function, and new approaches to therapy. *Advances in Pediatrics*, 63(1), 47–77. <https://doi.org/10.1016/j.yapd.2016.04.005>.
- Khan, M. J., Gerasimidis, K., Edwards, C. A., & Shaikh, M. G. (2016). Mechanisms of obesity in Prader-Willi syndrome. *Pediatric Obesity*, 13(1), 3–13. <https://doi.org/10.1111/ijpo.12177>.
- Kundert, D. K. (2008). Prader-Willi syndrome. *School Psychology Quarterly*, 23(2), 246–257.
- Lambert, J. M., Parikh, N., Stankiewicz, K. C., Houchins-Juarez, N. J., Morales, V. A., Sweeney, E. M., & Milam, M. E. (2019). Decreasing food stealing of child with Prader-Willi syndrome through function-based differential reinforcement. *Journal of Autism & Developmental Disorders*, 49(2), 721–728. <https://doi.org/10.1007/s10803-018-3747-y>.
- Lang, R., Didden, R., Machalicek, W., Rispoli, M., Sigafos, J., Lancioni, G., Mulloy, A., Register, A., Pierce, N., & Kang, S. (2010). Behavioral treatment of chronic skin-picking in individuals with developmental disabilities: A systematic review. *Research in Developmental Disabilities*, 31(2), 304–315. <https://doi.org/10.1016/j.ridd.2009.10.017>.
- Lowe-Greenlee, B. (2019). *Prader-Willi syndrome: Parent perceptions of school, professional, social, and informational support, and relations between support, child behavior, and stress [Unpublished doctoral dissertation]*. University of North Carolina.
- Maglieri, K. A., DeLeon, I. G., Rodriguez-Catter, V., & Sevin, B. M. (2000). Treatment of covert food stealing in an individual with Prader-Willi syndrome. *Journal of Applied Behavior Analysis*, 33(4), 615–618.
- McAllister, C. J., & Whittington, J. E. (2011). A short clinical overview of Prader-Willi syndrome. *Clinical Obesity*, 1(4–6), 184–188. <https://doi.org/10.1111/j.1758-8111.2011.00022.x>.
- Page, T. J., Finney, J. W., Parrish, J. M., & Iwata, B. A. (1983). Assessment and reduction of food stealing in Prader-Willi children. *Applied Research in Mental Retardation*, 4(3), 216–228.
- Pokorski, E. A., & Barton, E. E. (2020). A systematic review of the ethics of punishment-based procedures for young children with disabilities. *Remedial & Special Education*. <https://doi.org/10.1177/0741932520918859>.
- Prader-Willi Syndrome Association. (2020). *Prader-Willi syndrome: Transforming and saving lives*. <https://www.pwsausa.org/>
- Rigamonti, A. E., Bini, S., Grugni, G., Agosti, F., DeColt, A., Mallone, M., Cella, S. G., & Sartorio, A. (2014). Unexpectedly increased anorexigenic postprandial responses of ppy and glp-1 to fast ice cream consumption in adult patients with Prader-Willi syndrome. *Clinical Endocrinology*, 81(4), 542–550. <https://doi.org/10.1111/cen.12395>.
- Spendelow, J. S. (2011). Assessment of behavioral and psychiatric problems in people with Prader-Willi syndrome: A review of the literature. *Journal of Policy & Practice in Intellectual Disabilities*, 8(2), 104–112. <https://doi.org/10.1111/j.1741-1130.2011.00296.x>.
- Veltman, M. W. M., Thompson, R. J., Roberts, S. E., Thomas, N. S., Whittington, J., & Bolton, P. F. (2004). Prader-Willi syndrome: A study comparing deletion and uniparental disomy cases with reference to autism spectrum disorders. *European Child & Adolescent Psychiatry*, 13, 42–50.
- Whittington, J., & Holland, A. (2010). Neurobehavioral phenotype in Prader-Willi syndrome. *American Journal of Medical Genetics Part C: Seminars in Medical Genetics*, 154c(4), 438–447. <https://doi.org/10.1002/ajmg.c.30283>.
- Zyga, O., Russ, S. W., & Dimitropoulos, A. (2018). The PRETEND program: Evaluating the feasibility of a remote parent-training intervention with children with Prader-Willi syndrome. *American Journal of Intellectual & Developmental Disabilities*, 123(6), 574–584.