Unique Considerations of Prader-Willi Syndrome

11

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Prader-Willi syndrome (PWS) is a genetic, neurodevelopmental disorder, occurring in approximately 1 in 12,000–15,000 live births, that has specific physical, medical, and behavioral characteristics (Dykens & Cassidy, 1996; Milner et al., 2005; Wigren & Hansen, 2003). This syndrome has been reported in all races and ethnic groups, although it is reported disproportionately more in whites (Thompson, Butler, MacLean, Joseph, & Delancy, 1999). Approximately 70 % of PWS cases involve a deletion on the long arm of the paternally derived chromosome 15 [del 15 (q11–q130)], often including the whole Prader-Willi/Angelman Critical Region (PWACR; Cassidy et al., 1997). Most of the remaining cases of PWS (approximately 25 %) are attributed to maternal uniparental disomy, which results when both chromosome 15s are derived from the mother. A small proportion of cases (less than 5 %) is due to a mutation affecting the imprinting center. In these cases, the paternally derived PWACR is present but is not imprinted. Despite these differences in genetic etiology, all subtypes of PWS are the result of a non-expression of paternal genes in the PWACR (Whittington et al., 2002). Although the presence of multiple physical, medical, and behavioral characteristics suggests PWS, genetic testing is necessary to make a diagnosis (Whittington et al.).

In this chapter, we will review the clinical and behavioral features of PWS with a focus on (a) hyperphagia and associated food-related problem behavior, (b) physical activity levels, (c) self-injurious behavior (SIB) (e.g., skin picking), and (d) other problem behaviors (e.g., physical aggression) that interfere with quality of life. We will also discuss assessment and treatment of these behavior disorders with a focus on behavior analytic methods involving functional analysis methodology and function-based treatment. Finally, we will discuss some of the unique considerations for assessment and treatment of behavior disorders in PWS.

Clinical Features and Symptoms

Individuals with PWS have common physical characteristics including short stature, small hands and feet, narrow forehead, almond-shaped eyes, hypogonadism, hypopigmentation (i.e., fair skin and hair), and a small mouth with downturned corners (Dykens, 1999; Dykens & Cassidy, 1996). In addition to physical characteristics, the development of symptoms is quite unique in this syndrome. Infants with PWS show hypotonia (weak muscles), poor reflexes, and difficulty sucking, which often results in "failure to thrive" and the need for tube feeding (Dykens, 1999; Dykens & Cassidy, 1996; Dykens, Lee, & Roof, 2011).

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Infancy is followed by a very large increase in appetite (hyperphagia) between ages 1 and 6 years (Cassidy et al., 1992; Thompson et al., 1999), which continues throughout the lifespan (Dykens, Maxwell, Pantino, Kossler, & Roof, 2007). Finally, many individuals with PWS show global developmental delays (e.g., delayed language and motor development) by age 6 (Dykens & Cassidy, 1996), and approximately half are diagnosed in the mild to moderate range of mental retardation with an average IQ of 70 (range, 40–105; Dykens, Hodapp, Walsh, & Nash, 1992; Dykens & Shah, 2003; Thompson et al., 1999).

The most serious and pervasive concern for individuals with PWS and their families is hyperphagia (overeating), which is hypothesized to be due to abnormal functioning of the hypothalamus (Dykens & Cassidy, 1996). Hyperphagia coupled with reduced metabolic rates and low physical activity of individuals with PWS results in rapid weight gain and obesity (Dimitropoulos et al., 2000; Dykens, 1999). In fact, individuals with PWS are reported to have 40–50 % body fat (two to three times more than in normal individuals) and low levels of lean body mass with the heaviest amount of fat in the trunk region and limbs (Thompson et al., 1999). Morbid obesity is associated with serious health concerns including hypertension, diabetes mellitus, congestive heart failure, and sleep apnea (i.e., pauses in breathing during sleep; Butler, 1990; Butler et al., 2002). In addition, eating large amounts of food quickly may result in choking or gastric dilation or rupture (gastric necrosis; Dykens et al., 2011; McAllister, Whittington, & Holland, 2010; Thompson et al., 1999). Finally, individuals with PWS rarely vomit or report gastrointestinal pain, making it difficult to determine an emergency situation such as some of those listed above. Thus, without intervention, hyperphagia is extremely dangerous and is the major cause of premature death in this population (Einfeld et al., 2006).

Behavioral Disorders

Individuals with PWS engage in various behavior problems related to hyperphagia that include food-seeking behaviors such as hoarding or stealing food, eating inedible or unpalatable items (i.e., pica), and stealing money to purchase food. However, individuals with PWS have also been reported to engage in other problem behaviors that are unrelated to food, including (a) SIB, (b) sedentary behavior (i.e., low levels of physical activity), (c) ritualistic behaviors that are often described as "obsessive compulsive," and (d) other problem behavior (e.g., physical aggression, tantrums, and property destruction) that may or may not be related to gaining to access to food (Dykens & Cassidy, 1996; Dykens & Shah, 2003; Thompson et al., 1999).

Hyperphagia and Food-Seeking Behavior

Hyperphagia is defined as an insatiable and voracious appetite. In fact, studies have shown that individuals with PWS do not have normal satiety or feelings of fullness (e.g., Holland et al., 1993; Holland, Treasure, Coskeran, & Dallow, 1995; Lindgren et al., 2000; Zipf & Berntson, 1987). For example, Holland et al. (1995) provided individuals with and without PWS unlimited access to sandwich quarters and measured the amount of food consumed by each participant. Results showed that individuals with PWS consumed three times as many calories as those without PWS and reported a delay in satiety (i.e., did not report "feeling full" until 40 min after access to food and only after eating an excessive amount of food).

Given their insatiable appetite, it is not surprising that individuals with PWS are reported to steal and hoard food and consume inedible items (pica). The results of several large-scale surveys, questionnaires, and interviews have suggested that individuals with PWS engage in food stealing and unhealthy food consumption (i.e., consumption of unappealing or dangerous items such as pet food, frozen chicken, rotten food, and vitamins; Dykens, 2000; Greenswag, 1987; Holm & Pipes, 1976). Greenswag (1987) provided structured questionnaires to 275 caregivers of adults with PWS and found that the majority of the caregivers reported that the individual with PWS engaged in food stealing and hoarding, which oftentimes necessitated the need to "lock

up" all food in the home. In addition, several experimental studies have shown that when left alone in a room with food items (and told not to eat the items), individuals with PWS will consume the "prohibited items" (Maglieri, DeLeon, Rodriguez-Catter, & Sevin, 2000; Page, Finney, Parrish, & Iwata, 1983; Page, Stanley, Richman, Deal, & Iwata, 1983). Dykens (2000) conducted an interview to determine whether individuals with PWS would be willing to eat contaminated food, partially contaminated food, unusual food combinations (e.g., hot dog and whipped cream), or inappropriate items (e.g., pasta and paint). In the interview, pictures were used to present different desired foods in the above situations, and the participants were asked to report their willingness to eat the item. The authors found that participants with PWS were more likely than others with and without intellectual disabilities to report that they will (a) eat contaminated food, (b) consume inedible items, and (c) eat unusual food combinations. However, it is important to note that consumption of these items may be most likely to occur under conditions of restricted access to food.

Sedentary Behavior

In addition to food-related problem behavior, another behavior that contributes to the obesity and overall health of individuals with PWS is their reported sedentary behavior (Greenswag, 1987; Hodapp & Dykens, 1994; Nardella, Sulzbacher, & Worthington-Roberts, 1983; Rankin & Mattes, 1996). Nardella et al. (1983) reported that parents of children with PWS indicated that their children were lethargic and tired for no apparent reason. Eiholzer et al. (2003) used pedometers to measure daily activity of a group of individuals with PWS and a group of typically developing adults. The experimenters found significant differences between the groups in their average daily activity levels, with members of the PWS group, on average, more sedentary. In a study by van Mil et al. (2000), the authors evaluated the amount of energy expenditure during both rest and activities and found differences between individuals with PWS and a control group of individuals with similar heights and weights. Using basal metabolic rate (BMR) and average daily metabolic rate (ADMR) measures, the authors found that individuals with PWS expended significantly less energy than their matched pairs in the control group. These data suggest that whether or not individuals with PWS are more sedentary than their typically developing peers, they overall expend less energy while active and at rest, which puts them at a higher risk for obesity. Lower activity levels, as well as lower levels of energy expenditure, in individuals with PWS may contribute to health problems.

Self-Injurious Behavior

SIB is defined as any behavior directed toward one's own body that produces injury (Tate & Baroff, 1966). Various topographies of SIB have been reported to occur in individuals with PWS including hand biting, headbanging, hairpulling, and rectal digging (Symons, Butler, Sanders, Feurer, & Thompson, 1999). However, the most common form of SIB is skin picking, which is reported to occur in 65–97 % of individuals with PWS (Dykens, Cassidy, & King, 1999; Symons et al., 1999). Skin picking is defined as scratching, pulling, digging, or gouging one's own body (Lang et al., 2010). Individuals with PWS have been reported to mostly pick the front of legs, head, and arms (Symons et al., 1999). Obviously, there are several health risks associated with skin picking (and other forms of SIB) including contusions and lacerations, infection, and scarring (Ho & Dimitropoulos, 2010). In addition to health risks, these behaviors often interfere with learning, independence, and quality of life. For example, the appearance of visible sores or wounds may prevent individuals with PWS from forming new relationships. If wounds become infected, the infection may impede the individual from interacting with others, which can hinder both social and skill development by preventing access to other people and activities (e.g., school, work). Finally, SIB that results in contusions and lacerations often requires constant supervision that may result in decreased independence (e.g., placement in restrictive settings).

Obsessive-Compulsive Behavior

Some individuals with PWS are reported to have symptoms similar to those diagnosed with obsessive-compulsive disorder (OCD; Dykens & Shah, 2003). In fact, individuals with PWS tend to score in the clinically significant range on instruments that measure the extent and severity of obsessive and compulsive behaviors (e.g., Yale-Brown Obsessive Compulsive Scale; Dykens, Leckman, & Cassidy, 1996). Obsessive-compulsive behaviors in individuals with PWS include hoarding (e.g., paper, toiletries), ordering and arranging items by specific characteristics, questioning repeatedly, being concerned with exactness and sameness, and redoing things (e.g., rereading, rewriting) (Dykens & Cassidy, 1996; Dykens & Shah, 2003; Dykens et al., 2011). Dykens et al. (1996) reported a significant association between obsessive-compulsive behaviors and PWS, and Dimitropoulos, Feurer, Butler, and Thompson (2001) found that individuals with PWS show significantly higher levels of compulsive behaviors than individuals with Down syndrome and typically developing individuals. In addition, Dykens et al. (1996) found that these behaviors were time-consuming and distressful for a high percentage of individuals with PWS and their families.

Other Problem Behavior

Individuals with PWS are reported to engage in other problem behaviors such as tantrums, property destruction, and physical aggression. Dykens et al. (1999) conducted a survey of a large sample of individuals with PWS and found that 88 % engaged in tantrums (e.g., yelling, screaming, and noncompliance), 42 % engaged in property destruction (e.g., throwing objects, breaking objects, hitting walls), and 34 % engaged in physical aggression (e.g., hitting, kicking, or biting others). Although these behaviors may occur to gain access to food, they may also occur for other reasons (to gain access to other social and nonsocial reinforcers such as attention from others or escape from aversive situations; see a more detailed description of these possible functional reinforcers in the *Assessment and Treatment of Behavior Disorders* section below). As is the case with SIB, these problem behaviors are often stressful for the individual and their families and caregivers and may affect learning, independence, and overall quality of life. For example, individuals who engage in aggression and property destruction may be precluded from going to certain places (e.g., museums or the mall) or attending certain events (e.g., concerts) out of fear that the individual with PWS may destroy expensive items or harm others. In addition, if the individual with PWS requires constant supervision, caregivers may be precluded from engaging in preferred social events. Therefore, severe problem behavior may lead to outcomes such as reduced quality of care, increased caregiver turnover, and placement in restrictive settings.

Assessment and Treatment of Behavior Disorders

A common treatment for hyperphagia and other behavior problems displayed by individuals with PWS is pharmacological intervention. However, no consistently effective pharmacological strategies have been developed for the treatment of individuals with PWS (Dykens & Shah, 2003). Recently, several studies (e.g., Craig et al., 2006; Hoybye, Hilding, Jacobsson, & Thoren, 2003) have found that growth hormone (GH) therapy may be effective over the longer term in reducing body fat, increasing lean body mass, increasing height velocity, and increasing overall height in individuals with PWS; however, it has not been shown to have any effect on eating behavior (McAllister, Whittington, & Holland, 2010). It is important to note that side effects and risks associated with GH therapy should be considered (Dykens et al., 2011; Dykens & Shah 2003). The major focus of this section will be on environmental manipulations and behavioral assessment and treatment of behavior disorders in PWS.

General Weight Management

Individuals with PWS eat more, require less calories to maintain an appropriate weight, and engage in lower levels of physical activity (with less energy expenditure) than typically developing individuals. Common weight management strategies have emerged from research and case studies that are regarded as best practice and include (a) implementation of a low-calorie, individualized diet with vitamin and calcium supplements (as determined by a dietician/nutritionist), (b) frequent weigh-ins (at least weekly), (c) participation in exercise (approximately 30 min per day), and (d) environmental manipulations to restrict access to food (close supervision in all settings, locks on food cabinets and refrigerators) or money to purchase food (Dykens et al., 2007; Dykens & Cassidy, 1996; Dykens & Shah, 2003). Although these strategies tend to be rigorous and require a major lifestyle change for individuals with PWS and their families, these strategies have shown modest success in reducing weight and maintaining healthy eating habits of individuals with PWS.

Notwithstanding evidence that the above strategies have been shown to be somewhat effective, compliance with these strategies by individuals with PWS and their families may not occur. Thus, behavioral interventions involving antecedent- and consequent-based manipulations have been implemented to ensure compliance with weight management programs. These interventions have included (a) nutrition education (Holland et al., 1995; Mullins & Vogl-Maier, 1987; Pipes & Holm, 1973); (b) positive reinforcement in the form of earning preferred items and activities contingent upon compliance with the weight management program (e.g., prescribed diet, exercise, weight loss, and the absence of food stealing; Marshall, Elder, O'Bosky, & Liberman, 1979; Mullins & Vogl-Maier, 1987); (c) response cost (i.e., loss of items or activities) contingent upon the occurrence of noncompliance with the weight management program (Altman, Bondy, & Hirsch, 1978; Marshall et al., 1979; Thompson, Kodluboy, & Heston, 1980); (d) self-monitoring of weight, exercise, and caloric intake (Burke et al., 2011; Chambliss et al., 2011; Altman et al.); and (e) contingency contracting (Altman et al.).

For example, Marshall et al. (1979) showed that a treatment package implemented on an inpatient unit with four individuals diagnosed with PWS was effective for producing weight loss. The treatment package involved smaller meal portions, loss of meals contingent upon consumption of unapproved food (response cost), and contingent access to preferred activities for weight loss and the absence of consuming unapproved foods for 1 week (differential reinforcement of other behavior, DRO). In addition, Altman et al. (1978) showed that a combination of self-monitoring of daily caloric intake, weight, and exercise; nutritional education (i.e., participants were taught to determine caloric points for particular foods); contingency contracting in which individuals with PWS were provided a contract that outlined the dietary program and the consequences for compliance and noncompliance of the program and weight loss (i.e., earning access to preferred items and activities for compliance on a daily or weekly basis and withholding of these items and activities for noncompliance); and response cost (e.g., monetary fines, loss of privileges) for food stealing resulted in a decrease in weight for two participants. Furthermore, weight loss was maintained when the program was faded to only self-monitoring and reinforcers for weekly weight loss and exercise adherence.

Special Considerations for Weight Management

Although various strategies and intervention packages have been effective at weight management in PWS, research has suggested that special considerations may need to be taken into account for optimal effects of a weight management program. First, research has shown that the composition of a prescribed diet may affect the likelihood of compliance with the diet. Second, food stealing may need to be specifically targeted to reduce weight and maintain weight

loss. Third, specific interventions may need to be used to increase and maintain physical activity levels of individuals with PWS to result in better weight management.

Food Preferences and Diet

As mentioned above, dietary management and intervention must include a low-calorie diet; however, it is unclear how this diet should be composed to be most effective for adherence and weight loss. Early reports suggested that individuals with PWS were indiscriminate in their food preferences (Holm & Pipes, 1976; Pipes & Holm, 1973); however, several research studies have shown that individuals with PWS have a preference for some foods over others (e.g., Caldwell & Taylor, 1983; Fieldstone, Zipf, Schwartz, & Bernston, 1997; Rankin & Mattes, 1996). For example, research has suggested that individuals with PWS have preference with respect to the quality of food. That is, they prefer sweet foods over salty, sour, or plain foods (Caldwell & Taylor, 1983; Taylor & Caldwell, 1985); high-carbohydrate foods over high-protein foods; and high-protein foods over high-fat or lower calorie foods (Fieldstone et al., 1997).

In addition to quality, several studies have shown that other variables (e.g., amount or magnitude of food) may affect preference (e.g., Caldwell & Taylor, 1983; Glover, Maltzman, & Williams, 1996; Joseph, Egli, Koppekin, & Thompson, 2002). For example, Glover et al. (1996) showed that individuals with PWS prefer a smaller amount of high-preference food over a larger amount of low-preference food; however, when comparing a smaller amount of high-preference food to a larger amount of mixed-preference (mix of medium and low preference) food, individuals with PWS preferred the large amount of mixed-preference food. This latter pattern of choice was in contrast to control participants who continued to choose the smaller amounts of high-preference food. In addition, Joseph et al. (2002) showed that individuals with PWS were more likely to choose a larger quantity of food (three pieces) as compared to a smaller quantity of food (one piece), even when the delivery of the larger quantity was delayed for a small period of time (15, 30, and 60s). This pattern of choice was in contrast to the choices of obese control participants who chose the different food options on an almost equal number of choice opportunities. Based on the results of study 2 of Glover et al., however, it is possible that choice of larger quantity over smaller quantity is affected by the relative preference of foods (e.g., large quantities of *very* low preferred foods may not be chosen over small quantities of high preferred foods). In summary, previous research has suggested that the quality and quantity of food may affect food preferences for individuals with PWS, and an interaction of these variables may affect preference (e.g., quantity may affect preference when foods are similarly preferred but not when foods have large differences in preference).

Although the controlled laboratory studies mentioned above suggest certain global preferences for the population of individuals with PWS, it is likely that the provision of a larger number of food choices and different quantities of food would result in idiosyncratic preferences with respect to different foods and variables that may affect food choices. Thus, assessment of food preferences and the variables that may affect food preferences might be an important assessment package to determine individualized diets for individuals with PWS that may, in turn, increase dietary adherence and reduce food stealing (Rankin & Mattes, 1996). Depending on the results of this assessment for a particular individual, dietary options might involve (a) shifting preferences toward lower calorie foods that have the same (or similar) quality as preferred foods, (b) interspersing larger amounts of low quality (and most likely low-calorie foods) with very small amounts of high-quality foods (while keeping within the daily caloric intake), (c) providing small amounts of high-quality foods several times throughout the day, or (d) providing a small amount of high-quality food for consuming lower quality food throughout the day or at a specified period of time.

A final point regarding preferences and food consumption is related to reports that individuals with PWS consume inappropriate foods and other items. Given that quality of food is an important

variable for food choices among individuals with PWS, it is surprising that they have been reported to consume unpalatable foods or inedible items. However, it is possible that the consumption of these foods is due to an ongoing state of food restriction due to current weight management programs, which may establish these foods or items as sufficient when all other food is unavailable. If this is the case, then it is possible that providing individuals with PWS access to no-calorie or extremely low-calorie foods throughout the day might decrease the occurrence of food stealing or consumption of inappropriate items.

Food Stealing

Assessment and treatment of food stealing may be challenging in individuals with PWS because this behavior is often covert (i.e., it occurs in the absence of others; Page, Finney, et al., 1983; Page, Stanley, et al., 1983). Assessing the variables that may affect food stealing and determining the effectiveness of treatment on this behavior are only possible by observing the occurrence (or some product) of food stealing. Thus, a challenge to assessing and treating food stealing is how to measure food stealing when it occurs covertly. Therefore, several measurement procedures have been successfully employed for measuring covert food stealing of individuals with PWS and other intellectual and developmental disabilities (IDD) and include using a room with a one-way mirror for observation and scoring of food stealing (e.g., Page, Finney et al., 1983), videotaping (via hidden camera) and scoring the occurrence of food stealing at a later point in time (e.g., Ringdahl et al., 2002), and measuring the permanent product of food stealing (e.g., weighing food or counting the number of food objects before and after sessions; Maglieri et al., 2000; Page, Stanley et al., 1983).

Using one of the above measurement procedures to evaluate the occurrence of food stealing, several studies have demonstrated the effectiveness of behavioral interventions based on differential consequences including reinforcement and mild punishment (e.g., verbal reprimands, response cost). Page, Finney, et al. (1983) were the first to measure and directly intervene upon food stealing behavior of individuals with PWS. These experimenters recorded the occurrence of food stealing and consumption of prohibited food (via monitoring through a one-way observation window) by two children with PWS. The experimenters implemented a DRO procedure to reduce the occurrence of covert food stealing. That is, the observers watched into the session room from a one-way observation mirror and, contingent on the absence of food stealing for a prespecified interval, entered the session room and provided praise and a token. If the participant engaged in food stealing during a particular interval, observers entered the room at the end of that interval and told the participant that he would not get a token because he stole food. If the participant earned ten tokens during a session, the tokens could be traded at the end of the session for preferred items or a low-calorie snack. Results showed that the DRO procedure was effective for reducing food stealing, even as the DRO interval (i.e., the period of time in which participants had to refrain from food stealing to earn the reinforcer) was systematically increased. Although the DRO procedure was effective for reducing food stealing, procedures were not employed for generalization and maintenance, and the reduction in food stealing did not seem to generalize to other settings.

In an extension of the intervention by Page, Finney, et al. (1983), Page, Stanley, et al. (1983) showed the effectiveness of a behavioral intervention on the covert food stealing of an individual with PWS and evaluated a program for generalization of the effects to more naturalistic settings. In this study, the first phase involved evaluating the effects of a DRO token program on reducing food stealing in three hospital settings on an inpatient unit. During this phase, food theft was measured by baiting the session rooms with food items and taking inventory of these food items prior to and after the session. The DRO intervention involved earning a token (to be exchanged for low-calorie snacks at the end of the session) for the absence of food stealing for a prespecified period of time and a verbal reprimand for food stealing. This intervention was effective for reducing food stealing even when

the DRO interval was slowly increased to 15 min; however, generalization did not occur to settings in which the intervention was not implemented. In the next phase, the program was expanded throughout the day to include contingencies for exercise, changes in weight, and food stealing on the inpatient unit. Tokens were earned for weight loss of at least 1 lb at weigh-ins (conducted twice per week), and response cost (token fine) was implemented for food stealing on the unit or during scheduled sessions as conducted in phase I. Results showed that the participant had a decrease in weight during this phase. Finally, a similar program was implemented when the participant was transferred back to her group home. That is, the same contingencies for exercise, weight loss, and food stealing were implemented by group home staff. Specifically, the participant could earn tokens three times per day for the absence of food stealing. In addition, tokens were provided contingent upon exercise (e.g., walking, swimming, or stair climbing) and self-monitoring (written records of food consumed and graph of body weight). The results of this phase suggested that reductions in food theft and weight loss were generalized and maintained in a more naturalistic setting.

Although the results of these studies suggest that food stealing can be successfully measured, behavioral intervention can be used to decrease food stealing and reduce weight, and procedures can be employed to result in maintenance and generalization of the effects, most interventions and maintenance programs continue to involve continuous and close supervision of individuals with PWS, which is often difficult, if not impossible. To increase the likelihood of maintained effects, we need procedures that will result in maintenance of reductions of food stealing in the absence of continuous direct supervision (Maglieri et al., 2000). One such procedure involves stimulus control. Maglieri et al. (2000) measured the occurrence of food stealing by weighing food in a session room before and after each intervention session. The initial intervention involved the use of verbal reprimands delivered either within session (for each instance of food stealing) or at the end of session. Both interventions were effective in reducing food stealing. To increase the generalization and maintenance of low levels of food stealing, the experimenters used a stimulus control procedure, wherein they paired reprimands with a warning stimulus (i.e., they placed an orange sticker on containers with items that the participant was forbidden to consume and told the participant that she was not allowed to eat the foods in the container with the orange sticker). The warning stimulus was used to help the participant discriminate between permitted and prohibited foods during sessions. At the end of the session, the experimenter came into the room and weighed the prohibited food container. If there was a difference in weight (food was missing), then the experimenter provided a verbal reprimand. The intervention resulted in a decrease in food stealing from containers labeled with the warning stimulus. In addition, the authors evaluated whether this decrease in food stealing would generalize to a different situation. They placed the sticker on a refrigerator containing pudding cups and found that the participant did not engage in food stealing. In a similar study with an individual with IDD, Piazza, Hanley, and Fisher (1996) showed that pairing response interruption with a purple card resulted in a decrease and maintenance of low levels of pica in the presence of the purple card (even when response interruption was no longer implemented).

Exercise

Participation in exercise is particularly important for individuals with PWS. Exercise facilitates weight loss and decreases loss of lean body mass, which is especially important for the health of individuals with PWS. Common prescriptions for exercise in this population include at least 30 min per day of exercise with an individualized exercise regimen prescribed in conjunction with a physician and fitness coach. Common exercise programs include walking on a treadmill, biking on a stationary bike, or other ongoing cardiovascular activity (e.g., dancing).

Given the danger of morbid obesity in this population, it is surprising that very few systematic treatments have been employed to increase

physical activity levels of individuals with PWS. One example of an effective treatment for increased exercise of individuals with PWS was described by Caldwell, Taylor, and Bloom (1986) in which the authors evaluated the effect of providing preferred foods contingent upon completing all required exercise on a given day. Results showed that highly preferred foods were effective in increasing the activity level of 7 of the 11 participants with PWS. In addition to actual food, calories may be earned contingent upon compliance with an individualized, daily exercise regimen. For example, a small amount of calories could be provided for every *X* amount of calories expended for exercise on a particular day (e.g., 25 cal for every 100 cal expended; Caldwell et al., 1986).

Although food and calories may be an effective reinforcer for increasing and maintaining exercise, their use may seem counterproductive to the goals of weight loss and dietary management for this population. Thus, once increases in exercise have occurred, the provision of edibles or calories should be faded to social reinforcers (e.g., interaction from others, access to preferred items and activities). Another possible intervention that has been shown to be effective for increasing exercise in individuals with other IDD and typically developing individuals is to provide access to preferred items and activities (e.g., preferred TV shows, movies, music) while exercising (i.e., as long as the participant is engaged in the exercise activity at criterion levels) to increase the likelihood of engaging in the exercise behavior (Caouette & Reid, 1991; De Luca & Holborn, 1992; Flittering, Martin, Gramling, Cole, & Milan, 1988; Lancioni et al., 2003; Wysocki, Hall, Iwata, & Riordan, 1979).

Self-Injurious Behavior

Due to infection, SIB such as skin picking should result in immediate and routine care including topical creams and bandaging. Simple manipulations might include cutting fingernails and limiting time in the contexts in which the problem behavior occurs (Stokes & Luiselli, 2009). In addition to these general safety procedures, assessment procedures and interventions based on learning principles have shown to be the most effective for treating SIB (including skin picking; Iwata, 1994). Determination of the interventions most likely to be effective for decreasing SIB (as well as other topographies of problem behavior) is derived from conducting a functional analysis (Iwata, Dorsey, Slifer, Bauman, & Richman, 1982/1994) in which participants experience different conditions for the purpose of determining the situations under which SIB is most and least likely to occur. Specifically, a participant is exposed to multiple "test" conditions in which potential maintaining consequences (e.g., delivery of attention, escape from demands) are withheld and delivered for the occurrence of SIB. The levels of SIB in each test condition are compared to a control condition in which the same reinforcers are delivered independent of behavior. Test conditions in which high levels of SIB occur as compared to the control condition indicate that problem behavior occurs to access that reinforcer. The functional analysis allows clinicians to determine whether SIB is maintained by social positive reinforcement (in the form of attention or access to preferred items or activities), social negative reinforcement (in the form of access to escape from difficult or aversive situations such as work tasks), or automatic reinforcement (in the form of sensory stimulation or pain attenuation) (see Betz & Fisher, 2011; Hagopian, Dozier, Rooker, & Jones (2013); Iwata & Dozier, 2008; Iwata, Kahng, Wallace, & Lindberg, 2000; Neidert, Bayles, & Miller in this handbook for a review of functional analysis methodology). Once the function of problem behavior is identified via functional analysis, a function-based treatment may be used to decrease the occurrence of SIB. Function-based treatment strategies involve (a) no longer providing the functional reinforcer for problem behavior (extinction; EXT), (b) providing the functional reinforcer for engaging in an alternative behavior (differential reinforcement of alternative behavior, DRA) or for the absence of problem behavior (DRO), and (c) modifying antecedent conditions that evoke problem behavior for the purpose of

decreasing motivation to engage in problem behavior (e.g., noncontingent reinforcement, NCR). See Carr, Coriaty, and Dozier (2000); Iwata and Dozier (2008); and Hagopian et al. (2013) for a detailed review of function-based interventions.

In a review of behavioral treatments for skin picking in individuals with IDD, Lang et al. (2010) reported that treatment approaches have included various antecedent- and consequent-based interventions similar to those mentioned above (i.e., DRO, DRA, and NCR). Other treatments involve the use of protective equipment (e.g., helmet) or materials (e.g., bandages, gloves) to prevent the occurrence of skin picking, which can be placed on the individual following skin picking or worn at all times. Finally, several forms of punishment have been used to reduce the occurrence of skin picking. These procedures have included verbal reprimands, response interruption, and overcorrection (e.g., contingent exercise). It is important to note that after the seminal article on functional analysis methodology was published by Iwata and colleagues in 1982, published studies on the assessment and treatment of skin picking (as well as other self-injurious and severe problem behavior) have moved toward less intrusive procedures that are based on the function of behavior (Lang et al., 2010); however, mild punishment and prevention procedures are still used if reinforcement-based procedures are ineffective.

Several studies have suggested that skin picking is often maintained by automatic reinforcement and, like food stealing, occurs mostly in the absence of others (i.e., covertly; Didden, Korzilius, & Curfs, 2007). Given the parts of the body that are most often targeted for skin picking (legs and head), it is possible that individuals with PWS pick areas of the body that are not easily visible to others (Symons et al., 1999). Didden et al. (2007) conducted indirect assessments on the skin picking behavior of 119 individuals with PWS and hypothesized that the majority of individuals sampled had SIB maintained by nonsocial reinforcers (i.e., automatic reinforcement) because the behavior occurred mostly when they were alone. If skin picking is maintained by automatic reinforcement, it occurs either to access some form of sensory stimulation (Rincover, 1978) or to attenuate some painful stimulus (Miltenberger, 2005).

Due to the covert nature of skin picking (like food stealing), measurement of the occurrence of the behavior (for the purpose of assessment and treatment) is difficult. However, similar procedures have been suggested to those mentioned above for covert food stealing (i.e., observation via a one-way mirror, videotaping, and permanent product measures of SIB). For example, Iwata, Pace, Kissel, Nau, and Farber (1990) created the Self-Injury Trauma (SIT) scale to determine the occurrence of SIB by measuring tissue damage (a permanent product measure). The SIT scale is a rating scale that allows one to determine the location, type, number, and severity of tissue damage caused by SIB. Thus, the scale could be administered prior to, during, and after treatment to evaluate the effects of treatment on the occurrence of SIB.

To determine whether SIB is covert, a modified functional analysis could be conducted (with an observation procedure used to be able to measure possible covert behavior). In the modified functional analysis, two conditions could be implemented to assess for SIB maintained by automatic reinforcement. Both conditions would include a barren environment and no programmed consequences for engaging in SIB. However, one condition would involve the presence of another person (no interaction condition) and the other condition would involve the absence of others (alone condition). If higher levels of SIB occur in the alone condition as compared to the no interaction condition, then this would suggest that the behavior is covert. For example, Paisey and Whitney (1989) measured the occurrence of pica of a female adolescent with IDD under several conditions including an alone and a no interaction condition. Results of the assessment showed that the highest level of pica occurred in the alone condition, suggesting that her pica was covert. In a recent study, Toussaint and Tiger (2012) compared the levels of skin picking displayed by a 12-year-old boy with multiple diagnoses in an alone and no interaction condition. Results showed that much higher levels of skin picking

occurred in the alone condition, suggesting that his skin picking was covert.

Assessment and treatment of behavior maintained by automatic reinforcement is often difficult (regardless of whether it occurs overtly or covertly) because the automatic reinforcer(s) maintaining the problem behavior is not under the immediate control of the therapist (Vollmer, Marcus, & LeBlanc, 1994). One treatment that has shown to be effective for decreasing automatically reinforced problem behavior (including SIB) in individuals with IDD is NCR (DeLeon, Anders, Rodriguez-Catter, & Neidert, 2000; Roane, Kelly, & Fisher, 2003; Roscoe, Iwata, & Goh, 1998). NCR as a treatment for automatically reinforced problem behavior involves providing free access to preferred items and activities that might compete with the occurrence of SIB (see Carr et al., 2000; Tucker, Sigafoos, & Bushell, 1998; Vollmer & Borrero, 2009 for a review of NCR for treatment of problem behavior). When using NCR, preferred items are initially delivered continuously or on a dense reinforcement schedule. A competing items assessment (e.g., Shore, Iwata, DeLeon, Kahng, & Smith, 1997) may be conducted to determine items that are likely to be highly preferred (i.e., result in high levels of item engagement) and compete with the occurrence of the problem behavior (i.e., result in low levels of problem behavior). Shore et al. (1997) conducted a study on the SIB of three individuals with IDD. Functional analysis results suggested that the SIB of each participant was maintained by automatic reinforcement. Next, the authors compared the effects of NCR (i.e., continuous access to leisure items) to a baseline condition in which no items were available. The authors showed that when NCR was implemented, the participants' SIB reduced to near zero levels. Therefore, it is possible that during times in which an individual with PWS who engages in skin picking is in barren environments or alone, providing access to high-preference items and activities will compete with the occurrence of skin picking or other forms of SIB maintained by automatic reinforcement.

If SIB is covert, there are additional challenges for intervention when constant supervision cannot be provided. Thus, delayed contingencies (e.g., reprimands for tissue damage or reinforcers for the absence of tissue damage) may be one method for reducing SIB. Grace, Thompson, and Fisher (1996) conducted a study in which the occurrence of SIB by a young woman with PWS was measured via permanent product during three physical exams per day. During the exams, the nurses recorded the appearance of open skin, blood, and objects in the ears, eyes, or nose on a diagram of the body. Treatment involved the delivery of tokens for the absence of tissue damage (DRO) noted during the daily physical examinations, and tokens were exchanged for access to attention, preferred activities, and tangible items. Results showed that the intervention resulted in a significant decrease in SIB (i.e., the percentage of physical examinations with new injuries decreased). Recently, Toussaint and Tiger (2012) implemented an intervention that did not require product measures or continuous observation to determine the occurrence of covert SIB for the purpose of implementing contingencies to reduce the occurrence of the behavior. The experimenters used a variable momentary DRO (VMDRO) procedure in which reinforcers (i.e., a token and praise) were delivered contingent upon the absence of behavior at a particular moment (i.e., at the moment of brief observation). Tokens were exchanged for 30-s access to preferred videos following each session or at the end of the day. Results showed that the procedure was effective for decreasing skin picking even when the DRO interval was slowly increased to 5 min. This procedure may be preferred by caregivers because it does not involve continuous observation or product measures that may not detect some instances of SIB (i.e., those that do not result in tissue damage). It is important to note that it is possible that the brief checks that were conducted to determine whether skin picking was occurring at a particular moment would have been effective without the DRO intervention. That is, the mere presence of a therapist may have decreased skin picking. This is possible given that the behavior only occurred at high levels in the alone condition of the functional analysis, which may have been due to a history of punishment (e.g., verbal reprimand) for skin picking in the presence of others.

Obsessive-Compulsive Behavior

Few studies have evaluated the effects of interventions for treating obsessive-compulsive behaviors in individuals with PWS or other IDD. However, much research has been conducted on treatment for these behaviors in typically developing adults and children. The most common forms of treatment for these behaviors are Exposure and Response Prevention Therapy (a form of cognitive-behavioral therapy [CBT]) and pharmacological intervention (see Mancuso, Faro, Joshi, & Geller, 2010 for a review). Exposure and Response Prevention Therapy has been shown in numerous studies over the past three decades to be the most effective intervention for obsessive-compulsive behaviors in individuals with OCD (Abramowitz, Taylor, & McKay, 2005; Houghton, Saxon, Bradburn, Ricketts, & Hardy, 2010). This therapy involves exposing the participant to the aversive situation (e.g., dirty hands for a compulsive hand washer) and preventing the participant from engaging in the compulsive behavior. It is presumed that obsessive-compulsive behaviors are maintained by automatic negative reinforcement in the form of escape or avoidance of an aversive stimulus (i.e., fear or anxiety). However, it is unclear whether all obsessive-compulsive behaviors across all individuals, populations, and topographies have a similar function. Therefore, it may be important to first determine the function of obsessive-compulsive behaviors displayed by individuals with PWS for the purpose of designing effective treatment. In a recent study, Rodriguez, Thompson, Schlichenmeyer, and Stocco (2012) conducted a study aimed at reducing compulsive behaviors including arranging and ordering of furniture, completeness of tasks (e.g., insisting that drawers and doors stayed closed), and straightening items on shelves or in refrigerators of three children with IDD. The experimenters first conducted functional analyses and found that all three children's compulsive behavior was maintained by automatic reinforcement. Next, the experimenters compared the effects of (a) providing access to matched stimuli (i.e., stimuli that could be arranged and ordered in an appropriate manner and on a smaller scale) or competing stimuli (i.e., items or activities that were preferred and competed with the occurrence of the compulsive behavior in a competing items assessment), (b) matched stimuli with prompts to engage with the matched stimuli, and (c) matched stimuli plus responses blocking (i.e., the therapist physically blocked the child from engaging in the compulsive behavior) or matched stimuli plus product extinction (i.e., placing the object back in the original position contingent upon arranging and ordering). The results showed that response blocking or product extinction was necessary to decrease compulsive behaviors to low levels, and some participants required continued prompting and reinforcement for engaging with the continuously available matched stimuli. As discussed by the experimenters, it is important to provide individuals with an alternative activity to compete with the occurrence of the compulsive behavior in addition to implementing response blocking or extinction. It is possible that continuous access to potent reinforcers that are otherwise restricted may compete with the occurrence of obsessive-compulsive behaviors.

Other Behavior Problems

Although food-related problem behaviors are the most severe and common behavior problems in individuals with PWS, other behavior problems (e.g., property destruction, physical aggression, and temper tantrums) should be assessed and treated. Functional analysis methodology has been shown to be effective for determining the functions of numerous problem behaviors across various populations (see Hanley, Iwata, & McCord, 2003 for a review). Therefore, best practice involves the use of this methodology to determine an effective function-based treatment that is likely to be effective. As mentioned above, common function-based treatments such as (a) DRA (or functional communication [FCT]) in which the participant is taught to appropriately request the reinforcer and problem behavior no longer results in the reinforcer (extinction [EXT]; Carr & Durand, 1985; Tiger, Hanley, & Bruzek,

2008), (b) DRO in which reinforcers are provided for the absence of problem behavior (Mazaleski, Iwata, Vollmer, Zarcone, & Smith, 1993; Vollmer & Iwata, 1992), and (c) various antecedent manipulations such as NCR to decrease the motivation to engage in the problem behavior (Carr, Coriaty, Wilder et al., 2000; Smith & Iwata, 1997) have been shown to be very effective for reducing various problem behaviors. It is important to note that these behaviors may be high in frequency and severity, which may result in dangerous situations for the individual with PWS and his or her caregivers. Therefore, in addition to function-based interventions, certain safety precautions and crisis management procedures may be necessary to keep the individual with PWS and others safe.

Conclusions

PWS is a multifaceted syndrome that is associated with numerous health and behavioral concerns. It is likely that most individuals with PWS will require some type of supervision for weight management and behavior management. At least initially, environmental safeguards (e.g., locked cabinets and refrigerators, alarms, enhanced supervision) may be required to reduce calorie consumption. However, it is our recommendation that intervention programs also involve nutritional education, self-monitoring of caloric intake and physical activity, frequent weigh-ins, and individualized intervention programs that are based on assessment results for optimal effectiveness as well as maintenance and generalization of effects. For example, food preference assessments that include different types of foods and different variables that may affect food choices can be conducted with a particular individual to determine a dietary program that will be most effective for compliance and ultimate weight loss. In addition, physical activity should be increased by providing potent reinforcers (as determined by individualized preference assessments) for completion of individualized exercise programs. It is possible that food or calorie reinforcers are required, at least initially, to increase physical activity; however, these reinforcers should be faded over time and be replaced with nonedible reinforcers (e.g., attention, preferred items, and activities). Furthermore, functional analyses should be conducted to determine the conditions under which particular problem behaviors occur for an individual, and these results should be used to create an individualized behavior plan to increase appropriate behavior and decrease problem behavior to access these reinforcers. Finally, special considerations should be addressed for problem behaviors such as food stealing and SIB that may occur covertly.

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