

Parent Perceptions on Managing Food Related Behaviors of Children with Prader-Willi Syndrome: A Qualitative Discussion

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April 18, 2008

Abstract:

Individuals with Prader-Willi syndrome (PWS) are cognitively impaired and have several compulsive behaviors, the most distinctive of which is over-eating and obsession with food. All of these children display negative behaviors that are usually associated with food. Current literature discusses some behavioral strategies that can aid in controlling these behaviors. The goal of this study is to determine what parents perceive as the most effective strategies to help control negative behaviors. A secondary aim of this study is a needs assessment for the PWS clinic that is opening at Primary Children's Medical Center this September (2008).

Introduction:

Prader-Willi syndrome (PWS) is a well-recognized genetic condition caused by a deletion of the paternal region of chromosome 15q11-q13. This single microdeletion results in a spectrum of physical and behavioral characteristics in affected individuals. Most notable is the compulsive drive for food and obsession with eating. The urge to eat is physiological and overwhelming; it is difficult to control and requires constant vigilance. Consequently, individuals with Prader-Willi syndrome are at risk for developing obesity due to overeating and gorging on food. Obesity is further challenging because these children have a significant reduction of energy expenditure and lean body mass when compared to obese individuals

While children with this pervasive neurodevelopmental condition have their individual personalities, they usually display a range of negative behaviors. These behaviors include severe temper tantrums, anxiety, scratching or picking skin and scabs, obsessive compulsive and ritualistic behaviors (asking questions repeatedly, fixating on ideas or activities), insistence on routine, eating inedible or unusual food items and combinations, stubbornness, manipulation, labile mood, and at times, violence. These behaviors are constant and difficult for caregivers to manage, as some individuals begin food seeking and will eat unpalatable foods, such as dog food.

There have been many descriptions of the neurological dysfunctions in some individuals with PWS that might contribute to their characteristic decreased satiety. An investigation on the neural basis of the hyperphagia shows that after food intake, this population displays abnormal patterns of neural activation that normally represents satiety (Hinton et al. 2006). Other studies have confirmed structural defects in the hypothalamus of several individuals with PWS, preventing them from feeling full (Shapira et al., 2005). Even with this knowledge, most medications have been unsuccessful in suppressing the

appetite of individuals with PWS, with no apparent reason as to why they do not work (Butler et al 2006).

Many strategies have been proposed that can help to decrease negative behaviors in this population. They focus on the idea that individuals with developmental delay are highly sensitive to their environment and can benefit from having a structured, routine setting in which to live. Individuals with PWS are characterized as having decreased ability to be flexible in their environment; therefore, it is quite beneficial to have rules, expectations, predictability and consistency to minimize frustrations and anxiety. This suggests that using control and routine in daily rituals would minimize negative behavioral traits.

According to the literature (Butler et al. 2006), there are 4 main methods to control food-related behavioral modifications for individuals with PWS: (1) structured food environment, (2) diet and exercise plan, (3) planning time and menus for each meal, and (4) denying all other access to food. Further, a study by Joseph et al (2002) showed that individuals with PWS chose larger quantities of food delivered after a delay in time, rather than smaller quantities of food given immediately. This demonstrates that larger portions of food are more desirable in order to fully satiate the individual with PWS.

Despite a vast description of behavioral methods that can be used to modify the food intake and behaviors of affected individuals, few studies have been conducted to determine the best methods for controlling negative behaviors of affected individuals. In this study, we created questions related to diet and behaviors and developed themes from interviews with parents or caregivers of children with PWS that demonstrate methods used that best help impact negative behaviors.

As part of the study, we also conducted a needs assessment by asking parents what services would be most useful for them in a clinic setting. The Primary Children's Medical Center is in the process of developing a Prader-Willi syndrome clinic, and having this information could greatly add to its success.

Methods:

For this study, I interviewed parents or caregivers of children who have PWS. Participants were drawn from the active PWS parent support group in Salt Lake City, Utah. I wrote a letter, approved by the University of Utah IRB, describing my study and asking for participants. The president of the PWS support group sent this letter out to all the families listed in the group (N=68). People who were interested in participating sent me an email and we arranged a time to set up an interview. The parents or caregivers could only have children between the ages of 5 and 18 years. I was looking both for individuals with PWS who have a deletion of the region on chromosome 15 and those who have uniparental disomy.

The interviews were about 1 hour each. They took place either in the families' home or in an otherwise specified meeting place, whatever was convenient for the

individual. During the interview I recorded the session with an ipod and a microphone. Questions were asked in three categories: diet, behaviors and clinic needs assessment. The diet plan/management were developed to determine the structure of the child's food environment. The behavioral questions focused on specific behaviors and how they related to the environment. Finally, clinic needs assessment questions were targeted to determine if the parents had health care providers help them formulate diets or behavioral interventions for their child.

Results:

A total of 14 interviews were conducted. Eleven interviews were recorded and listened to at least three times to sort out themes. Each time, parts of the interview were transcribed. Later, the quoted material was categorized into one of 6 themes based on similar word choice or concepts presented in the statement. One interview stopped recording early into the session, and two others were not recorded due to human error. Notes were taken during all interviews; therefore, this information was categorized for the 3 interviews that were not recorded.

Six major themes were identified from the transcribed interviews. Four of these themes were previously reported in the literature and include control, routine, portion size and positive reinforcement. Two new themes were identified from these interviews, and they include being strict and manipulating food.

Control was the most common theme reported by every participant. This theme was divided into 3 subthemes: planning/setting expectations, stable environment, and helping prepare food. Many parents reported that making plans for meals or other activities helped to ease anxiety and negative behaviors in their child:

“I think the behavior problems stem from anticipation...or thinking that she's not gonna get something, she wants to know what it's gonna be...if we don't have a plan that's when we get a meltdown”

Keeping a stable environment is another previously proposed strategy that is thought to lessen anxiety and increase positive behaviors in these children. Every participant in this study reported that their children's behaviors were worse when out of their home environment:

“If she's out of her environment, she'll ask for food more...once the food's put away it's ok, but if the foods out and she's not in her environment, it's a focus”

Finally, allowing the child to help prepare a meal was reported by several families as a way to alleviate negative behaviors. These parents believe that allowing the child to participate in preparation of the food gives them control over the situation, and thus decreases their anxiety. This method was not previously reported in the literature.

The next most common theme that was identified in this study was routine. This was split into two subthemes: having a set schedule and serving the same meals. Participants relayed that having a fixed schedule for meal times was essential for their child and helped to decrease anxiety:

“He’s very compulsive about his schedule, I don’t think he necessarily consciously knows it, it’s just, that’s what’s ingrained into him...he’s a creature of habit”

Routine also appeared as a theme when parents described serving the same meals to their child for breakfast or lunch. With this method, the children know what to expect for the meal and therefore they do not get anxious about what they will be eating. All participants described feeding their child the same breakfast every day. Further, if there were options available, the child would usually pick the food they normally have rather than the new meal available:

“Even if I make pancakes or waffles... she wants to have the cereal, she knows she’s gonna like, it’s her favorite cereal”

Many families described using portion size as a way to increase their child’s satisfaction with their meals. The subthemes for this category were using smaller plates/presentation of food, and giving a larger amount of healthy food. As described earlier, it has been shown that these children prefer a larger quantity of food at each meal. Many parents used plates that were smaller and filled them with food to make it appear as if they had a larger quantity of food than everyone else:

“We use small plates so I kind of look at portion size on dinner more than the actual calories...I mean the portions are small but it looks like there’s quite a bit of food there based on the size of the plate”

Other parents use plates that are the same size as everyone else’s, but they present the food in a way that makes it look like a bigger portion than it is:

“We squish out the food to make it look bigger, like it’s more food”

Further, every participant reported giving their child large amounts of healthy food rather than small amounts of unhealthy food. Some families give larger portions of salad or vegetables at each meal, while others give their child more than 5 healthy items in each meal to give variety:

“If she can have three times more carrots than she can of something else that we’re having, she’ll take more carrots than a little tiny bit of something else...there are choices, most of the time she’ll take the choice that she can have more of”

Positive reinforcement was the next theme that was previously reported in the literature as a method to control behaviors. In this study, only 2 participants reported using this strategy to perpetuate desired behaviors. A subtheme of this category was identified as reinforcing healthy eating. This allows children to feel good about eating healthy and be able to deny food at times, reducing their anxiety over wanting to eat:

“The neighbor wanted to give her a cookie, she said ‘I’ll just have to see what it is...I don’t eat unhealthy food’”

The theme of being strict emerged as a new strategy for decreasing negative behaviors and anxiety. Several parents mentioned that using punishments and keeping them consistent were effective methods for controlling negative behaviors. Therefore, the two subthemes were punishments and consistency. In one instance, the punishment of the child involved food, and in other situations it involved limiting an activity:

“If you pick, you have to have a slimfast for dinner, and we don’t bend that rule at all...he doesn’t bawl about it or cry, he knows. It’s been an effective behavior tool for him”

Many participants reported that being firm and keeping punishments consistent was another way of decreasing negative behaviors:

“On the behavioral end of it, if she just knows you mean business and if you’re firm then she’ll be ok, just that consistency and not allowing her to get away when you’re trying to keep the behavior in check”

Finally, many parents manipulate food in an effort to minimize behaviors and appease their children. The subthemes were categorized as making a plate or food special, using food as a reward or using tricks with food. Some parents reported that they give their child a special plate; the plate may be decorated or somehow different from everyone else’s in some way, but it is also smaller than the rest of the plates. However, the child does not know this and thinks instead that he is getting a special plate.

“So, he now is at the point where he has a very nice plate. Same color, matches everyone else’s, but it’s slightly smaller...he doesn’t know that, he thinks he gets a special dinner plate so he feels like he’s getting a special treat”

A few participants use food as a reward for chores or for other desired behaviors. This strategy has been reported previously in the literature; however, only two families reported using this method of behavioral control. Some parents discussed how food is a huge motivator for these children, but it can get them into trouble at times. However, the families that do use food as a reward say that their child tried harder to control behaviors in order to obtain the reward, and they know if they’ve behaved poorly that they won’t receive it.

“If he behaves good all week at home and school, he can buy hot lunch at school on Friday”

Finally, almost every participant reported using tricks with food in order to alleviate negative behaviors. These tricks mainly include creating the illusion of larger amounts of food in order to appease the child:

“If we have pizza, she wants two slices, we just take one piece and cut it in half. She watches us do it, she just doesn’t get it”

Other examples included giving the child many little items of food in a meal in order to make it seem like a large meal.

A variety of clinicians and providers were reported as being desired for a PWS clinic. Approximately half of the participants had help from dieticians or nutritionists in formulating a diet. One participant utilized a local neurobehavioral clinic in helping control her daughter’s behaviors, which were quite violent.

Discussion:

The aim of this study was to determine what behavioral methods parents were using most consistently to lessen negative behaviors in their child with PWS. Controlling the child’s environment and keeping a firm schedule for meals and activities were the most helpful methods to control behaviors and anxiety. However, even with set meal times for their child, several children asked repeatedly about meal times. In those situations, parents would help the child determine the amount of time left to wait and then distract them with another activity before the meal. Many parents report that when their child is out of their environment, these and other behaviors are significantly worse. Further, the behaviors are not limited to issues with food, as parents relayed outbursts during parties, exercises or at school. Overall, families use a variety of methods to help control the child’s environment and keep their lives routine because this is consistent with what their children want.

Families who met with a dietician or who used diet books initially counted calories for each meal. However, only three families in this study reported counting calories for meals, and even then dinner was not included as one of those meals. Reasons for this included the difficulty in determining calories for each type of food, which resulted in a lack of variety in meals, and many families did not have time to count calories for each meal. Instead, most parents utilized smaller portion sizes during meals to monitor food intake. This also allowed the child to eat the same meals as the rest of the family, eliminating the aspect of coveting other food items. Further, many parents offer their child larger portions of healthy foods. We know from previous studies that these individuals are more satisfied when given a larger quantity of food. Therefore, by giving ample amounts of healthy food, the child is receiving a low calorie diet, but is happier with the amount of food they are receiving and are less likely to act out.

Positive reinforcement has been previously reported to be a powerful method of perpetuating desired behaviors in these children. However, only two families described using this technique in the management of their child's negative behaviors. It's possible that more specific questions to target this theme would have yielded higher responses amongst individuals. Instead, the subtheme of reinforcing healthy foods was described by several families. This enabled their child to feel good about refusing unhealthy foods and decreased anxiety over desiring foods.

Several families in this study described the new theme of being strict and initiating punishments if their child continuously displays negative behaviors. Punishments either included being denied food or a being denied a favorite activity due to inappropriate behaviors (such as skin picking or tantrums). Current descriptions of behavioral management in textbooks do indicate that having a stringent regimen for limiting access to food will inevitably stabilize behaviors over time. However, there is no discussion of initiating punishments to help control these behaviors. This strategy is even more appealing because by allowing parents to give punishments, it enables them to normalize their child's behaviors and act on them as they would for an unaffected child.

Finally, manipulating food was a method that many parents used to preemptively alleviate negative behaviors. This strategy was not previously discussed in texts that describe strategies for behavioral management; however, almost every family reported having special treats for their child or using tricks with food to create the illusion of larger portion size, for example. These methods enable the child to feel good about what they are eating and feel special, thereby eliminating the desire for other food items. Using food as a reward for appropriate behaviors was only reported by two families in this study. Many families discussed that food can be an excellent motivator, but should be used with caution.

The limitations of this study include a small sample size and a specific group of individuals from Utah. However, since this is a qualitative study, there is less need for a larger sample size. For more quantitative data, it would be preferable to have more participants.

It is well known that PWS impacts not only the entire family, but also individuals at school, friends of the family, and the community itself. For many families, the initial diagnosis is a struggle between denial of the situation and dealing with the numerous health problems present in their child. These struggles change over time, and families find themselves dealing with significant behavioral problems in their child. Many parents have said that the behavioral problems are more difficult to manage than the multitude of health problems. Therefore, an intervention for controlling behaviors should be implemented as early as possible into the child's life.

The strategies described in this study are a good starting point for recommendations on methods to control negative behaviors. Geneticists at the PWS clinic opening at Primary Children's Medical Center are planning to include these recommendations for behavioral

control when seeing patients. Based on information obtained from this study, these include:

1. Having a structured and routine food environment
2. Setting up expectations for food or other activities
3. Utilizing smaller portion sizes or variety for meals
4. Manipulating food to create the illusion of a special treat or larger quantity of food
5. Enforcing punishments and keeping them consistent

These methods should be started as early as possible in order to have the child in a routine. Our goal is to help families control these behaviors as well as for the children themselves to be comfortable in their environment.

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