

Characterizing and Managing Behavior in Prader-Willi Syndrome

Kevin Jackson, Ph.D., CBA
ARC of Alachua County
Gainesville, Florida

This series of articles is based on my training as a behavior analyst and on my thirteen years of experience in working with children and adults with PWS in family settings, schools, and as director of behavioral services for a large residential treatment program specializing in PWS. The concepts and interventions discussed here have been extremely effective in addressing the behavior of individuals with Prader-Willi Syndrome.

PART I. ABOUT BEHAVIOR

The occurrence of behavior is influenced and controlled by the environment.¹ The environment includes both the physical environment and the social environment. The phone rings and we answer it. The temperature drops and we go for our jackets. We get a letter from an old friend and we spend the day thinking about them and remembering. Another friend fails to return our calls and we quit calling them. A child with PWS notices that activity materials in the classroom have been put away in the wrong place and becomes upset and argumentative when not allowed to correct the problem. In fact, the environment is so important to behavior that there is an entire science dedicated to studying the ways in which environmental factors control behavior. That science, behavior analysis, is a thriving part of the larger discipline of psychology. Although most of psychology focuses on questions of how the mind (or brain) might be organized, or how thinking relates to behavior, behavior analysis is concerned directly with how the environment affects behavior (including the behavior of thinking), and on developing methods for arranging the environment to influence behavior in beneficial ways. This is important from a practical point of view because it is much easier to manipulate the environment than to manipulate someone's brain or mind.

The Logic of Dealing with Behavior

Behavior is controlled by the environment.

We can control the environment.

Therefore, we can control behavior.

¹ The emphasis on environmental control is not meant to imply that physiology is not also relevant to behavior. Physiology and environment are both relevant to most all instances of behavior, especially with Prader-Willi syndrome. Just as a physiologist may write of physiological control over behavior without discussing the possible role of the environment, these articles are written by a behavior analyst or behaviorologist from the perspective of environmental control without going into the possible role of physiology or neurology. The present set of articles are designed to highlight some basic environmentally based practices of immediate practical benefit in working with individuals with Prader-Willi Syndrome.

Basic Rules of Behavior

1. Consequences strengthen or weaken behavior. That is, what happens after some behavior occurs determines the future probability or strength of that behavior. You go to a new shoe store, wait thirty minutes for assistance, and the sales person is rude. These consequences will likely weaken the behavior of going to that particular store in the future. In the hallway at school, on her way to the restroom, a girl with PWS notices some food in a trashcan and eats it. Searching through trashcans while on bathroom breaks becomes a regular part of the girl's school routine. In this case, food reinforces the behavior of searching through trashcans.

Consequences that strengthen behavior are called reinforcers (reinforcement), and consequences that weaken behavior are called punishers (punishment). It is important to realize that punishment, as described here, does not necessarily correspond to what we mean by punishment in everyday language. In fact, everyday examples of punishment may not actually weaken behavior at all. In some cases, consequences administered as punishment may actually strengthen the very behavior they are intended to decrease. Similarly, rewards for appropriate behavior might not actually strengthen, or reinforce, the behavior.

2. Immediate consequences are more powerful than delayed consequences. What happens immediately or shortly following the occurrence of behavior is generally more powerful in determining the occurrence of that behavior, than what happens after some delay. For example, while shopping an individual might come across an expensive item (reinforcer) that they would like to own. If the person does not have enough money to purchase the item immediately, the person is faced with a choice between two behaviors. First, the person could save up the money or wait until the necessary money is available. With this choice, the person would get the item, but only after some delay. The second option is to pay for the item with a credit card and get the item immediately (often at a higher cost in the long run). Using the credit card is often more likely to occur because it produces more immediate reinforcement. The profitability of the credit card industry relies on this characteristic of human behavior.

Immediate consequences are also more powerful when it comes to problem behaviors. A parent may be puzzled when, despite consistently losing privileges at home for having tantrums at school, a PWS child continues to disrupt the classroom with tantrums. However, it is likely that the more immediate consequences of tantrums at school are more important than anything a parent can do hours later at home.

3. Behavior that occurs today is often a result of the consequences that the behavior produced in the past. The restaurant you choose to go to today depends on the past consequences of going to that restaurant and others. Similarly, whether a child with PWS complies with a parent's request, or is noncompliant with that request, depends on what the consequences of compliance and noncompliance have been in the past.

4. The occurrence of behavior at a given time may be triggered by events that occur

immediately before that behavior. For example, demanding bossy requests may be more likely to trigger noncompliance, verbal resistance, or tantrums. Friendly, nonthreatening requests may be more likely to result in compliance. Similarly, in individuals with PWS, a prompt to switch from the current activity to another, being told no, and disruptions of the usual routine, often trigger problem behaviors.

What about Genetics?

Understandably, it might cause confusion to emphasize the environment when talking about behavior in PWS. After all, one might ask, aren't the behaviors associated with PWS a result of genetics?

A useful way to think about genetics is that genes determine how the environment will affect the individual. For example, suppose there is a gene linked to an increased risk of certain forms of cancer. Rather than directly causing a person to get cancer, the gene may simply increase the probability of cancer when the individual is exposed to certain environmental carcinogens. That is, the gene may be viewed as determining the effect of environmental cancer causing agents. In the absence of gene therapy treatment, such a person would be especially wise to avoid known carcinogens.

Similarly, when it comes to behavior, genes do not directly cause behavior but determine how the environment affects behavior. Genes do not directly cause tantrums in PWS. They increase the chances that certain environmental circumstances will result in tantrums. In the absence of effective gene therapy, it is important to understand some of the ways environmental variables characteristically, or uniquely, influence the behavior of individuals with PWS. Such an understanding can help us to more effectively arrange environments to prevent behavior problems and promote appropriate and beneficial behaviors.

Consider the behaviors of excessive eating, or food seeking, in PWS. Genetic differences do not directly cause the PWS individual to remove the hinges from a locked food-containing cabinet, or to dig in a trash can for food debris. Rather, the genetic defect characteristic of PWS appears to result in food being a strong reinforcer even when the individual has already eaten some quantity of food. Obviously, food is a reinforcer for all of us, but typically its reinforcing value is diminished as we consume food and only returns after some extended period of time without food. If food is almost always a reinforcer in PWS, any behavior that results in access to food, or increases the chances of getting food, will be reinforced and will be likely to occur again and again. This includes raiding the kitchen after others are asleep, digging through trashcans, or volunteering to pass out snacks at school. In our program, we take advantage of this susceptibility to food as reinforcement. For example, we have shown that daily, intense, aerobic exercise can be established and maintained on a long-term basis when food is delivered as a consequence of exercise participation.

Other behavior problems associated with PWS can also be viewed in terms of differences in the way the environment influences the individual. Consider oppositional behavior,

noncompliance, arguing, and tantrums. These behaviors are characteristic of PWS. What are the typical consequences of these behaviors? That is, how are these behaviors typically responded to by others? Could social consequences from others be strengthening these problem behaviors and increasing their chance of occurrence? It appears so. Social disapproval, reprimands, and angry emotional responses from others, which typically follow many of the disruptive behaviors characteristic of PWS, often appear to directly reinforce the behavior. The following example illustrates this point.

While attending the YAAP at the national PWS conference in San Diego, I volunteered with a colleague to watch after a 10-year-old girl with PWS who was continually sneaking off and trying to run away from the program. Her behavior, no doubt, resulted in demands to return, warnings that she could get in trouble, and disapproval from others. She was crying when handed over to us. The first thing we did was to escort her to an area with few exits. When she did try to leave, we simply got in the way. We physically blocked her from leaving without getting angry, asking her to stop, or prompting her in any other way. After two or three attempts she was slowing down and looking for other options. We tried unsuccessfully to initiate a conversation with her. There was a barrier wall at the back of this area, overlooking the hotel entrance below. As she approached this wall, I held my hand to my heart and yelled out emotionally, "STOP! Get away from that wall! That is too scary, you will give me a heart attack." With her hand on the wall, she turned her head toward me and smiled. It was a large full smile from ear to ear. Her eyes were wide and her face lit up as if it were Christmas morning. She turned back around, raising herself to look over the wall again. I ran to her, continuing my insistence that she step back from the wall. She smiled, holding the wall, and said she wasn't afraid. She repeated the behavior many times as I continued to protest. She spoke with us on occasion and answered our questions about her name and where she was from. She never tried to leave again. She found a new behavior, with a bigger pay-off (consequence). I became more upset by her looking over the wall than by her running away. She quit running away and spent the rest of her time hanging out by the wall. Occasionally, she would look back at me to see my reaction.

If social disapproval and negative emotional reactions from others are reinforcers in PWS, then this information provides us with a mechanism for minimizing many behavior problems. In part, we can do this by refusing to deliver these typical consequences following disruptive behaviors. We have dozens of anecdotal demonstrations of this effect. For example, both the Pittsburgh center and our treatment program have found that skin-picking increases when the PWS individual is reprimanded as a consequence of this behavior. We usually recommend that minor skin picking behavior be ignored.

In conclusion, I have some good news about behavior and PWS. Like most people's behavior, the behavior of individuals with PWS can be reinforced by positive emotional social responses from others. That is, approval, praise, excitement, applause, and hugs all appear to be powerful reinforcers. In the next article I will begin describing how you can take advantage of these reinforcers as I introduce the five most important things you can do to improve the behavior of your child with PWS.

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PART II. IMPORTANT THINGS YOU CAN DO TO IMPROVE THE BEHAVIOR OF YOUR CHILD WITH PWS

Parents can learn to deal more effectively with the behavior of their child with PWS while learning and practicing parenting skills that are relevant to all of their children. This is true because the same basic parenting practices that are most effective with ordinary children are also most effective with PWS. Unfortunately, parenting practices which are ineffective or counterproductive with ordinary children, are especially harmful and counterproductive with PWS. For this reason, I believe it is critical for parents of a child with PWS to learn how to parent more effectively. Such parents will need to spend more time and energy dealing with problem behaviors than the parents of a non-PWS child. However, by practicing effective parenting techniques, such as those described below, parents can significantly decrease the overall amount of time and energy they spend dealing with problem behaviors.

1. Focus on Positive Interactions

Interactions between people may be categorized as positive or negative:

Positive Interactions consist of smiles, greetings, compliments, friendly gestures, eye contact, supportive gestures (e.g., thumbs up, o. k. sign), small talk, expressions of care and concern (e.g., "let me help you with that"), enthusiasm, and friendly, nonthreatening and nonjudgmental advice.

Negative Interactions consist of criticism, frowns, arguing, screaming, threatening verbally or through gestures and actions, talking down to or lecturing in the traditional parental way, showing disapproval, scolding, and bossy or demanding talk.

A social environment consisting of predominately negative interactions directly contributes to problem behaviors. Conversely, a predominately positive social environment can reduce or eliminate many problem behaviors. In a healthy social environment, positive interactions outnumber negative interactions by a ratio of at least eight to one. That is, there should be at least eight times as many positive interactions as negative interactions. We know that people are happier, healthier, more cooperative, more productive, and less likely to exhibit problem behaviors when their social environment is a positive one.

If you think that a ratio of eight to one is not attainable, I can assure you that it is. Consider infants in their first year of life. They are typically smothered with love and affection (i.e., positive interactions) probably at a ratio of one hundred to one or greater. Recently, I watched in amazement as my daughter's track coach ran an entire meet, with about twenty-

five girls participating, while remaining focused on positive interactions. In fact, he had a kind word for each and every participant in every single event. Let me tell you, those girls were motivated, well behaved, and they had a great time! Studies with children in family settings show that, when positive interactions sufficiently outnumber negative interactions, the children are more likely to be successful in school and to be emotionally, intellectually, and socially advantaged.

Some people, and some families, naturally focus on positive interactions. However, most of us have room for improvement in this area. I recommend taking your own data as a method of enhancing your own interaction ratio. Use a sheet of paper or 3x5 card with two columns labeled for positive interactions and negative interactions. Make a mark in the appropriate column for each interaction you have with your child (or children). You will find that the more positive interactions you engage in, the fewer problem behaviors your child will exhibit. Occasionally, I get called into a school because of the emergence of persistent behavior problems by a child with PWS. The first thing I do is observe the interaction ratio. It is often the case, that the teachers and staff have come to rely primarily on negative interactions in dealing with the child. When sufficient numbers of positive interactions are introduced, the problem behaviors typically decrease or disappear completely.

One key to creating a positive social environment is to eliminate coercion. Coercion means using criticism, sarcasm, threats, arguing, questioning, and/or physical or verbal force to control behavior. Eliminating coercion does not mean abandoning discipline or letting your child get away with murder. It means replacing primitive disciplinary practices with practices that actually lead to long-term beneficial behavior change. Coercion may appear to work because sometimes a coercive interaction can stop an ongoing problem behavior. However, this short-term effect is misleading. In actuality, the overall probability of the child engaging in problem behaviors can be expected to increase with the continued use of coercion. This is true, in part, because coercion motivates the individual being coerced to retaliate, get back at, and get even with, the people and social situations providing, or associated with, the coercion. Noncompliance, resistance, tantrums, theft, and aggression often occur as forms of retaliation in response to coercion by others.

In addition, coercive practices contribute to problem behaviors by directly demonstrating inappropriate behavior to the child. The coercive emotional outburst of an angry parent serves as a model of unacceptable behavior. This behavior is readily imitated when, at a later time, the child is angered by some event. For this reason, when it becomes necessary to intervene with problem behaviors, it is best if the parent is in control, and the consequences are administered calmly and confidently.

Coercion may be particularly harmful for children with PWS. As described in the last article, the behavior of children with PWS often appears to be directly reinforced by the emotional coercive responses of others. Coercion may also be more likely to trigger an emotional tantrum in a child with PWS. You will find that by staying focused on positive interactions, and by learning the other interventions I describe, you can comfortably and more effectively control your child's behavior without resorting to the use of negative or coercive interactions.

2. Ignore Systematically

There is a natural tendency for people to ignore appropriate behavior and to attend to inappropriate behavior. However, studies of social practices in education, business, and family settings show that attending to appropriate behavior while ignoring inappropriate behavior is a more effective option. Not only is ignoring a safe alternative to coercion, it is also one of the most potent options for eliminating problem behaviors. Ignoring means refraining from social interaction (e.g., eye contact, talk, gestures, and facial expressions) when a problem behavior occurs. It means pretending you never saw or heard the behavior and are going on as if it didn't occur, or as if you completely don't care.

A. Ignore Harmless Annoying Behavior

Ignore behaviors that do not threaten the basic quality of life, limb, and property. Annoying noises, talk, and silly or mildly disruptive activities all fit in this category. Pick your battles. Do not waste time and energy on minor behavior issues that are inconsequential. Instead, focus on your child's appropriate behaviors by attending to them with praise and positive interactions. Learn to tolerate and ignore the harmless behavior of your child with PWS. This includes age typical behaviors characteristic of all children, minor PWS typical behaviors (e.g., skin picking), as well as other peculiar behaviors that may annoy you but are actually harmless. For example, brief episodes of yelling or mildly aggressive play should be ignored. Unusual noises or verbalizations, which may be fun for your child but annoying to you, should be ignored. Minor instances of noncompliance, when immediate compliance is not essential, should be ignored. With noncompliance, a good strategy is to wait patiently for compliance and then reinforce compliant behavior, when it occurs, with a positive interaction. If a problem behavior is so annoying that you cannot stay and ignore, leave the area. You can go outside, go to another room, or walk away and do something else. If you absolutely cannot ignore the behavior, try redirecting the child to another area or activity with a friendly, non-threatening prompt. In either case, wait for your child to do a more appropriate behavior and attend to this behavior instead.

B. Ignore Arguing

Individuals with PWS are often predisposed to argue. Therefore, it is especially important to address arguing as a problem behavior. First, you can prevent some arguments by not saying "no" to your child's requests unless you are sure that you mean it and there is a good reason to say "no". Sometimes parents say "no" to a request initially (perhaps because it is not a convenient time or the request requires some effort on the part of the parent) and then after further argument and consideration give in and say "yes". This practice may inadvertently reinforce arguing. The next time the parent says "no" the child will be more likely to argue the point. In general, you should honor your children's requests unless there is a good reason not to. If you say "no" to a request, then stick to it. If saying "no" results in an argument, ignore it.

There also is a distinction between calmly asserting one's position about an issue and aggressively arguing. If your child is calmly discussing an issue with you, it is fine to listen and consider their perspective. If your child engages in loud, aggressive, or persistent arguing, ignore it.

The following outlines a basic strategy for addressing argumentative behavior in PWS. When used consistently, this strategy works remarkably well.

1. Make your decision about the issue and verbally state your conclusion.
2. If possible, have your child repeat back your decision and praise this response.
3. If you have an opportunity, briefly redirect your child to some other activity or situation. (For example, don't you need to feed your lizard now?)
4. Completely ignore any additional arguing or discussion on the topic.

C. Ignore inappropriate behavior that occurs in the context of appropriate behavior.

This is the most sophisticated form of ignoring. It usually involves ignoring inappropriate talk while attending to other appropriate behavior. The following scenario illustrates the technique:

The following interaction takes place prior to leaving on a shopping trip.

Mom: "Oh Tommy, that shirt doesn't fit. How about going back to your room and putting on another one that fits better."

Tommy: (while slowly walking back to his room)

"But it does fit mom. . . . Who cares anyhow, it's my shirt, not yours, I am sick of this, it is not fair!"

Mom: Says nothing and walks away to do other activities. She completely ignores Tommy's inappropriate talk.

(Tommy is doing exactly what his mother asked. His argument is harmless behavior that should be completely ignored.)

(Five minutes later, Tommy returns with another shirt)

Mom: noticing Tommy's shirt, approaches Tommy and gives him a hug, "Thank you Tommy, that shirt really looks much better. Now we can go to the store."

(Here, Mom initiates a positive interaction based on Tommy returning with an appropriate

shirt. She is reinforcing his compliant behavior, while ignoring his minor inappropriate arguing. An additional reinforcer in this scenario is the opportunity to go to the store, which was withheld by the mother until Tommy complied with her request by changing his shirt.)

If the mother is consistent in the use of this technique, Tommy's arguing will virtually disappear. Mom has removed the payoff for arguing.

The interventions described here are presented confidently. They are based on good science and research. I use these techniques myself, and I have seen them work time and again. This does not mean that implementing these methods is easy. Learning to do things differently requires considerable work and effort. It requires a commitment and a plan. To help, a parenting book I recommend is The Power of Positive Parenting by Dr. Glenn Latham. This book may be ordered at 1-800-748-4850 or from the web site (parentingprescriptions.com) where related materials are also available.