



Prader-Willi
California Foundation

Supporting People with Prader-Willi Syndrome

**Prader-Willi Syndrome
Behavior Management
Training Session**

PRADER-WILLI CALIFORNIA FOUNDATION
LISA GRAZIANO, M.A., LMFT, EXECUTIVE DIRECTOR

PRADER-WILLI SYNDROME

Even before 1956 when Prader-Willi syndrome was named by Swiss physicians Andrea Prader (Prah-der), Alex Labhart, and Heinrich Willi, families have struggled with how to manage behavior that is common amongst people diagnosed with PWS.

This presentation will provide an understanding of how PWS impacts the brain and the body and how we as parents, extended family, teachers, and care providers can develop strategies to better manage the behavior of our loved one, student, or patient.

No matter what the age of your individual with PWS you can reduce the number of unwanted behavior incidents when you understand how PWS impacts the individual and make the appropriate accommodations and changes in their environment.

PRADER-WILLI SYNDROME IS A SPECTRUM DISORDER

Symptoms of PWS vary in degree and severity
from person to person despite the
sameness of PWS type
(Deletion / UPD / Imprinting Defect)

We don't yet fully understand the physiology of PWS, but part of the problem lies in the Hypothalamus, the part of the brain that controls the release of hormones that regulate:

- Growth
- Muscle Tone
- Breathing
- Sleep Cycles
- Temperature Regulation
- Metabolism
- Emotions
- Hunger and Satiety

Physiological Symptoms

- Hypotonia - Weak Muscle Tone
- Abnormal Growth (short stature, small hands & feet)
- Problems with Strength, Balance, Coordination, Motor Planning
- Hyperphagia - Dysfunctional Appetite Regulating System
- Respiratory Issues
- Gastrointerological Issues–Gastroparesis, slow bowel motility, inability to vomit
- Hyper- & Hypothermia - Irregularities in Body Temperature Regulating Systems
- Incomplete Sexual Development
- Hypopigmentation
- High Pain Threshold
- Disordered Sleep
- Cognitive Limitations
- Speech and Language Issues (Dyspraxia)
- Dental Problems (decreased saliva production can cause severe problems). Biotene toothpaste helps!
- Skin Scratching and Picking
- Temperament and Behavior Issues with older children through adulthood
- Social / Psychological / Psychiatric Problems
- Other common characteristics include, but are not limited to: scoliosis, eye abnormalities, medication sensitivity, orthopedic problems

PARENTAL AND CARE PROVIDER STRESS

Parents raising a child with PWS face extraordinary stressors. Each feels the weight of the world and the weight of their family on their shoulders.

Research ([Hodapp RM](#), [Dykens EM](#), [Masino LL](#), 1997) shows that “compared to reported stress levels in families of children with mixed etiologies of retardation, parents of children with Prader-Willi syndrome showed higher levels of parent and family problems, and comparable levels of pessimism.”



STRESS



- It's imperative that parents and care providers find and create ways to manage their stress:
- Look to each other for support
- Use the Tag Team Approach
- Use stress reducing breathing techniques
- Exercise
- Girlfriend time / Guy time / Alone time / and especially Couple time

PWS CAN BE CONSIDERED A TWO STAGE DISORDER

Stage I: Failure to Thrive

- Hypotonia (muscle weakness)
- Feeding problems
- Respiratory problems
- Delayed developmental milestones



Hypotonia continues throughout lifetime and can contribute to behavior problems.

- Decreased strength and stamina
- Fine and gross motor skills deficits
- Daytime sleepiness
- Impaired balance and coordination

Growth Hormone therapy at any age can improve hypotonia and the resulting challenges

HYPERPHAGIA CONTINUED

- The brain of someone with PWS functions as if it believes the body is starving:
 - It drives the individual to eat as much as possible
 - It stores food as fat
 - It lowers the metabolic rate to about half conserve energy
- The brain does not receive and/or process signals of feeling “full” and drives the individual to get more food.
- Unmanaged hyperphagia + slowed metabolic rate causes rapid weight gain and potential for morbid obesity.

FACTORS THAT COMPLICATE HYPERPHAGIA

- Absent vomit reflex. The body does not expel consumed poisons, rotten food, too much food. Syrup of Epicac may *not* induce vomiting; repeated doses may be toxic.
- Delayed stomach emptying (gastroparesis)
- Slow emptying bowel (Miralax etc)
- Increased risk for sudden death from just one food binge (stomach rupture, tissue necrosis, poisoning)
- Choking is one of the top causes of death

TREATMENT OF HYPERPHAGIA

- No genuine learning to control the hyperphagia food drive.
- No medication – yet – to successfully reduce the drive to eat.
- Researchers working to develop a medication or medical intervention that will reduce/eliminate PWS’s hyperphagia symptom. (We are hopeful that Qsymia, now approved by the FDA, will prove successful. Qsymia is a combination of two older drugs that have long been known to help with weight loss: phentermine (the safer half of the banned fen-phen drug) and topiramate.
- “Treatment” of hyperphagia consists of 24/7/365/lifetime oversight of the individual with PWS and restricting access to all food sources.

HOW DOES A PERSON WITH PWS THINK?

Most individuals have decreased intellectual and adaptive functioning skills.

Average IQ typically 55-70.

General ranges of IQ distribution:

- 5% IQ 85+
- 30% Borderline Intellectual Disability
- 35% Mild Intellectual Disability
- 25% Moderate Intellectual Disability
- 5% Severe Intellectual Disability



DSM-5 scores Intellectual Disability based upon adaptive functioning and IQ scores rather than IQ test scores alone

PWS THINKING CONTINUED...

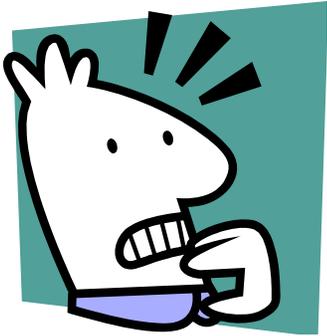
- **Oppositional–Defiant tendencies.** The brain’s automatic reaction is often to respond with “NO!” This looks like oppositional, defiant, argumentative behavior. If you say ‘black’ the person with PWS will typically say ‘white.’
- **Egocentrism.** People with PWS generally think about themselves first and foremost: *It’s All About Me.*
- **Impulsivity.** Persons with PWS generally want what they want, when they want it, which is now.
- **Obsessive–Compulsive tendencies.** The brain believes ‘if 1 is good, 100 is better.’ Collecting, hoarding, picking, sorting, pulling, tearing can be associated with the brain’s obsessive/compulsive tendencies.

PWS THINKING CONTINUED...

- **Low tolerance for frustration.** Anything that looks, sounds, or smells like a frustrator could lead to a behavior problem.
- **Inflexibility.** Persons with PWS tend to be inflexible.
- **Self-Monitoring.** The ability to monitor and evaluate one's own performance is often impaired.
- **Planning, organizing, prioritizing, and problem solving** skills typically poor.
- **Need to be correct.** Many people have a strong need to be right at all costs.
- **Short-term / working memory** is often poor.
- **Long-term memory** is usually excellent (thanks to the hormone ghrelin) so that once something is learned, it's remembered forever.

PWS THINKING CONTINUED

- Higher cognitive abilities and verbal acuity do not equate to ability to manage themselves around food.
- The ability to plan and execute complicated schemes to obtain food does not generalize to the ability to perform complicated tasks in other areas.
- Learning is not usually generalized from one situation to the next. What is learned in one context may need to be re-taught in another similar but different context.
- Persons with PWS are generally concrete thinkers. Terms like, “Hop to it!” may not be understood to mean “Begin the task immediately” and may cause confusion, anxiety, and result in an unwanted behavior.



PWS IS ALL ABOUT ANXIETY

Persons with Prader-Willi syndrome typically feel high levels of anxiety – all of the time.

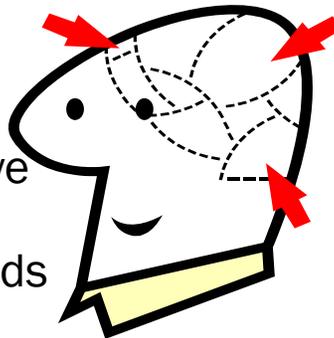
Maladaptive behaviors may be attempts to reduce the level of anxiety felt:

- Excessive talking
- Repeated questions
- Skin picking
- Arguing
- Controlling behavior
- Oppositional / defiant behavior

PWS IMPACT UPON EMOTIONS & BEHAVIOR

PWS Causes HYPER-REACTIVITY!

People with PWS generally have great difficulty managing their emotions. Whatever is felt tends to be felt with *great* intensity.



**Care Providers Must
Always Respond Calmly**

Cognitive impairment, impaired sensory-integration, slower processing speed, speech and language problems make understanding the environment more difficult and raises the level of anxiety.

Increased anxiety leads to more rigid thoughts in an attempt to make sense of the world and reduce anxiety.

More rigid thoughts lead to inflexibly clinging to current behavior in order to bring order and reduce anxiety.

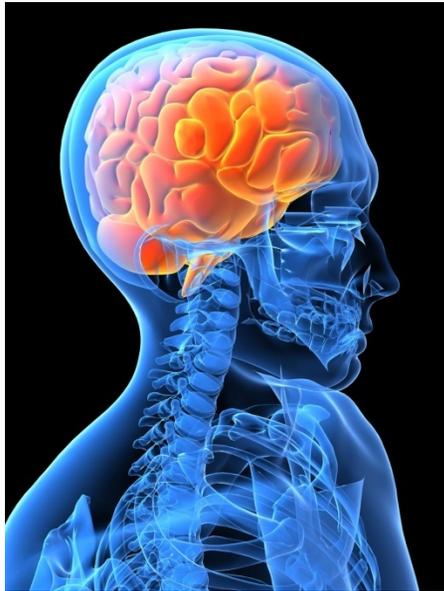
More rigid behavior is frequently problematic and interferes with the flow of the day.

MILLION DOLLAR QUESTION:

How do you manage
unwanted behaviors and meltdowns?

FIVE CENT ANSWER:

Avoid them in the first place!



**WE CAN'T CHANGE THE BRAIN
IN OUR LOVED ONE, STUDENT,
OR PATIENT WITH PWS**

**WE CAN CHANGE OUR OWN
BEHAVIOR WHICH *DOES*
CHANGE THE BEHAVIOR OF
THE INDIVIDUAL WITH PWS**

IDENTIFY POTENTIAL STRESSORS

- There is *always* some precipitating event to or reason for a behavioral incident, even if you have no idea what it is, even if it appears that the meltdown came “out of nowhere” or even if the reason doesn’t make any sense to you.
- Think ahead to identify potential stressors or situations that may cause anxiety or be misinterpreted or present some “chance” that may lead to a disappointment.

**Identify/eliminate/reduce the cause of anxiety
and you'll reduce the potential for a behavior problem!**

THINK LIKE THE PERSON WITH PWS

- **Oppositionalism:** Avoid “Yes/No” questions; Offer preferred choices; Give as much control as possible; Allow lots of time to process past the brain’s oppositionalism; Build in extra time everywhere you go.
- **Egocentrism:** Don’t say, “If we don’t hurry Grandma will be late to her appointment.” Do say, “If we get Grandma to her appointment on time you and I will have time to watch a movie tonight!”
- **Impulsivity:** Prepare before entering stores. Reward for patience.
- **Self-Monitoring:** Continuously remind to speak softly; slow down eating.
- **Need to be Right:** Providers responses: “Interesting...”; “You might be right.”
- **Planning, Prioritizing & Problem Solving:** Continuously provide assistance.
- **Obsessions:** Give preferred choices. Use empathy. Meds may be helpful.



BREAK

The ABC's of PWS Behavior Management

- Create Consistent Routines
- Create Clear Rules
- Create Clear Boundaries
- Create a Calm Environment
- Follow the Principles of Food Security
- Apply the Principles of Food Security to *every* area of life to create the Principles of Life Security

↓ Anxiety + ↑ Boundaries + ↑ Consistency
= ↓ Behavior Problems!

CREATE CONSISTENT ROUTINES



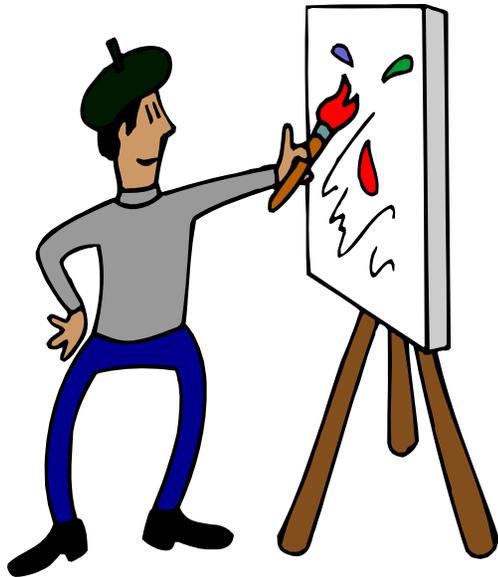
Consistency and routines help people feel safe and secure. When people know what to expect they don't have to worry about it. Routines reduce anxiety.

People with PWS have a strong need for routine, sameness and consistency.

Create routines around wake up time, meals and snacks, chores, bedtime rituals... *everything!*

- Visual calendars / schedules / auditory alarms are helpful.
- Transition cues – verbal countdown prompts, visual countdowns such as timers – are often necessary.
- Advance notice about changes in plans or routines can be helpful.

Paint the Picture



Before entering a new or unknown situation paint the picture of what things will look like, what will happen. Talk about your expectations for behavior.

- Knowing what to expect will reduce anxiety
- Fix a past goof before making another

Note: Giving too much advance notice may create anxiety. Not providing enough advance notice may create anxiety. Finding the right timing is important... and difficult.



CREATE CLEAR RULES

- People with PWS are rule followers. Rules provide understanding for what is expected and how to behave. If something is understood and processed as a rule or a contract, it will be adhered to.
- Establish household rules (make bed in the morning), hygiene rules (wash hands after using restroom), chores rules (set the table before supper), exercise rules (walk first then snack), social rules (greetings, sharing, turn-taking, reciprocal play), shopping rules (no whining), and food rules (Principles of Food Security).
- Be sure to establish the rule that parents can change a rule if necessary!
- Parents need to be parents – not friends - so their kids can feel safe. Best parental match for a person with PWS is one who is an authority figure but NOT authoritarian.

CREATE CLEAR BOUNDARIES



- Establish boundaries around what behavior is acceptable and what behavior is not acceptable.
- Say what you mean and mean what you say. Don't threaten something you don't intend to follow through on.
- Give limited, all preferential choices. "Do you want to wear the red jacket or the blue jacket?" "Do you want to take your bath/shower in 5 minutes or in 10 minutes?"
- Lead like a Mamma Duck. Don't wait for person with PWS to initiate movement.
- Be clear, avoid ambiguity. "We'll see" or "Maybe later" are too vague and may create anxiety which could lead to an unwanted behavior.



CREATE A CALM ENVIRONMENT

- Persons with PWS are typically “Hyper-Reactive.” The best environment for someone with PWS is one where everyone responds calmly.
- **Always speak calmly** during emotionally charged exchanges. The global sense of anxiety people with PWS feel is significantly increased by raised, critical or angry-sounding voices. Tone is often more critical than the words used.
- Respond calmly on the outside, despite how you feel on the inside. Be patient. Be gently firm. Be encouraging.

CALM ENVIRONMENT CONTINUED

- **Don't argue.** Don't engage in a power struggle. Use the refrain, "You might be right." It's ok to matter-of-factly explain, "It's this or nothing. What would you like?" Allow the individual lots of time to process their situation and work through their oppositionalism.
- **Don't try to talk someone out of their upset because they'll just feel they need to express *more* upset so that you understand.** The time it takes to listen and express genuine empathy is worth it. Listen, repeat their concern. Listen, repeat their concern. Listen, express *your* concern. Listen....
- Some people with PWS like to "stir the pot" to provoke a reaction. When they elicit a negative reaction, however, they feel anxiety which can turn into an unwanted behavior. Respond calmly to diffuse hyper-reactivity.
- **Respond to an escalation with indifference, boredom.** Ignore unwanted behaviors as much as possible to extinguish them as quickly as possible.



funny fuse

Don't Give In to a Tantrum. Don't Give In to a Tantrum.

If you give in to a tantrum you have taught the person with PWS all they need to do to get what they want is to cry louder, wait you out longer and that eventually you will give in.

Don't Give In to a Tantrum. Don't Give In to a Tantrum.

UH OH. ALREADY GIVEN IN TO A TANTRUM?

**Don't be too hard on yourself.
We all goof every now and again!**

- Explain that your past giving in has not been helpful.
- Apologize for your foible.
- Explain that from now on, for the child's/adult's benefit, you won't give in and you'll work harder to keep him/her safe.
- When a tantrum for something begins, calmly remind the individual that you will not be giving in because that's unfair to him/her.
- Stay strong and don't give in!

IMPACT OF FOOD MANAGEMENT ON BEHAVIOR



Many of the behavioral problems typical of persons with PWS can be reduced by instituting the **Principles of Food Security***

**Term coined by Janice Forster, M.D. and Linda Gourash, M.D.
of The Pittsburgh Partnership Specialists in PWS*

THE PRINCIPLES OF FOOD SECURITY*

NO DOUBT

No doubt when the next meal or snack will be served

+

NO CHANCE or HOPE

No chance or hope to obtain unauthorized food

=

NO DISAPPOINTMENT

That unauthorized food is not available

NO DOUBT

NO DOUBT WHEN THE NEXT MEAL OR SNACK WILL BE SERVED

- Establish consistent routines for food consumption. Determine when food will be served (every 2 ½-3 hours)

Breakfast / Morning Snack / Lunch / Afternoon Snack / Dinner

- Establish consistent rules for food consumption
 - Serve food on smaller size plate.
 - Cut food in small pieces, spread out to fill entire plate.
 - Baggie of snack cut or cracked into multiple pieces looks like more.

**Scheduled food should be like breathing air:
NEVER withheld for poor behavior and NEVER used as a bribe**

NO CHANCE OR HOPE

NO CHANCE OR HOPE TO OBTAIN UNAUTHORIZED FOOD



- If food is available, the individual can do little else but think of ways to get that food. Access to food creates hope. Hope creates anxiety. Anxiety creates the potential for unwanted behaviors. Eliminating access to food reduces behavior problems.
- Restrict access to food by using locks on the refrigerator, food pantry, kitchen, medicine cabinet, liquor cabinet, etc.
- Establish rules for type, quantity of food especially prior to attending parties, restaurants, etc.
 - No family-style bowls on table.
 - No second helpings unless you ALWAYS do seconds.
- Put a lock on the thought that unauthorized food is available.

NEVER Give In to a Tantrum for food.

NO DISAPPOINTMENT
THAT UNAUTHORIZED FOOD IS NOT AVAILABLE

No disappointment means fewer behavior problems

Life is good when there's no disappointment!

FOOD SECURITY TIPS

- Even the most honest person with PWS may lie and/or steal when it comes to food. This behavior is a symptom of PWS.
- If it is suspected that unauthorized food has been eaten do not question the food consumption (i.e., “Did you eat those chips?”). Rather, presume the food has been eaten and state as a fact (i.e., “I see you ate the chips.”). Reduce calories in future meals to compensate.
- Do not remove food from the hands of person (unless dangerous or poisonous) as this will create a behavioral outburst.

PWS Food Pyramid

Sweets
Carbs &
Starches

Fruits

Nuts

Dairy

Protein

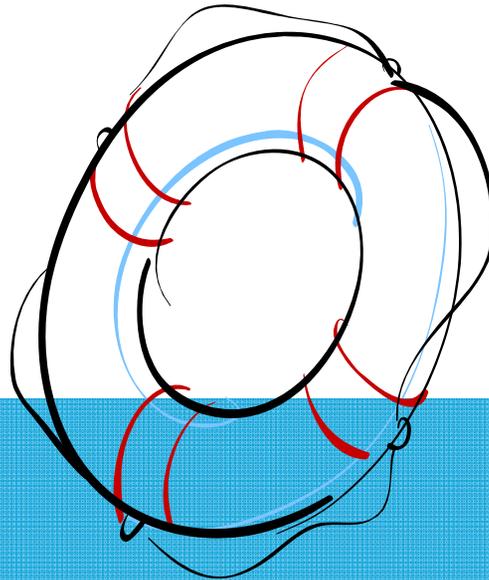
Vegetables

Dinner Plate



The Principles of Life Security

Apply the Principles of Food Security to
ALL PARTS of LIFE
to reduce anxiety and unwanted behaviors



THE PRINCIPLES OF LIFE SECURITY

- Create No Doubt/No Chance regarding **Rules**
- Create No Doubt/No Chance regarding **Routines**
- Create No Doubt/No Chance regarding **Boundaries**
- Create No Doubt/No Chance regarding a **Calm Environment**
- Implement the **Principles of Food Security**



BREAK

COLLABORATIVE PROBLEM SOLVING

Developed by Ross Greene, PhD to help manage the behavior of persons who are prone to react impulsively and explosively.

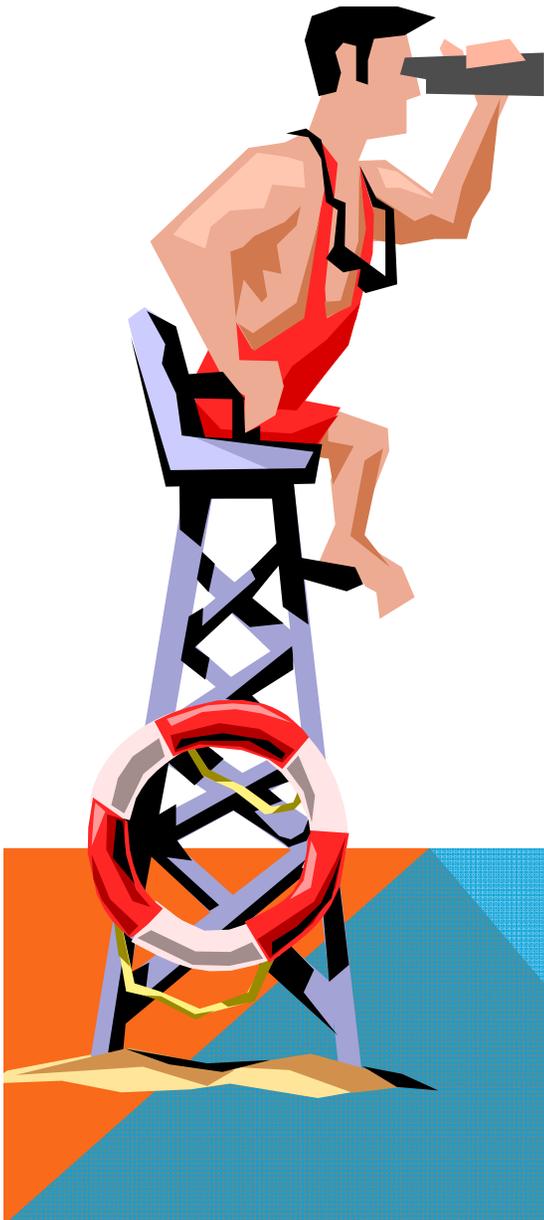
Book Referral: *The Explosive Child: A New Approach for Understanding and Parenting Easily Frustrated, Chronically Inflexible Children*

Center for Collaborative Problem Solving: www.ccps.info

“Instead of asking yourself, ‘What's it going to take to motivate this kid to behave differently?’ ask ‘Why is this so hard for this child? What's getting in his way? How can I help?’” ~Dr. Ross Greene



COLLABORATIVE PROBLEM SOLVING



Look for the potential compromise in order to *avoid* the meltdown, and *teach* the person with PWS to better develop their frontal lobe skills.

Dr. Greene believes caretakers too frequently behave *as if* something is uncompromisable when there really *could* be a solution that avoids a meltdown and gets both parties what they want.

EXAMPLE OF A COLLABORATIVELY-SOLVED PROCESS

Care Provider: It's just about is time to take a shower.

Person with PWS: No! I don't want to take a shower.

Care Provider: Oh, you don't want to take a shower yet. What's your concern?

Person with PWS: I'm not finished yet!

Care Provider: Oh, you're not finished yet. Is your concern that you don't want to take a shower until you're finished coloring?

Person with PWS: I don't want to take a shower until I'm all done!

Care Provider: I understand. Your concern is you want to finish coloring before your shower. My concern is it's getting late and you'll be too tired tomorrow to enjoy your outing. How do you think we can we work this out?

Person with PWS: I know! I can color 10 more minutes and then take a shower.

Care Provider: That's brilliant! I'll set the timer for 10 minutes. Thank you for working it out with me!

COLLABORATIVE PROBLEM SOLVING

“The hardest part about doing Basket B is remembering to do Basket B.”



CONTRACT AGREEMENTS

Use PWS's concrete thinking to your advantage by utilizing contract agreements.

- Anything written is more likely to be followed.
- Contract agreements help each party understand the other's concerns.
- Contract agreements can be created about *anything*.



A SUCCESSFULLY WRITTEN CONTRACT:

- Is focused on the *process* of the agreement
- Is always mutually created
- Keeps the individual with PWS thinking
- Incorporates the oppositional nature of PWS
- Pulls ideas and compromise from both parties
- Incorporates praise throughout the process
- Maintains calmness throughout the process
- Incorporates the Principles of Food Security
- Incorporates the Principles of Life Security
- Incorporates the Principles of Collaborative Problem Solving
- Incorporates patience, respect and neutrality throughout the entire process

SAMPLE CONTRACT AGREEMENT FORM

Contract Agreement Form

Only individuals who are mature and responsible enough may enter into a contract agreement. Does the individual believe he/she is mature and responsible enough to enter into a contract?

| | |
|---|-------|
| What does the Individual want? What is the concern of the Individual? | |
| | |
| What is the Parent's/Care Provider's concern about what the Individual wants? | |
| | |
| How can you work it out? Brainstorm possible solutions. | |
| | |
| Write down agreed upon solution(s). Be specific and detailed. | |
| | |
| Individual's Signature: | Date: |
| Parent/Care Provider's Signature: | Date: |

NEGATIVE REINFORCEMENT AND PUNISHMENT

People tend to want to use punishment for unwanted behavior.
Traditional behavior plans tend to emphasize negative consequences.

Examples of negative consequences include:

- Spanking
- Coercion (i.e. threats of losing a privilege)
- Termination of participation/services
- Taking something away (short or long term)



REASONS WE RATIONALIZE USING NEGATIVE CONSEQUENCES

- “There must be some consequence for her actions or she’ll just think she can do it again.”
- “What will the others think if she gets away with that?”
- “He will never learn what is right or wrong without being held accountable.”

WHY ARE NEGATIVE CONSEQUENCES NOT EFFECTIVE FOR PEOPLE WITH PWS?



- To learn from consequences a person needs:
 - Insight and ability to problem solve
 - Memory, logic, and ability to rationally build on past experiences
 - Ability to compare and discriminate information
 - Ability to recognize the value of an experience and learn from it
 - Ability to think sequentially (first this, then that; if this, then that)

PWS compromises or impairs most of these requisit skills

MORE REASONS WHY NEGATIVE CONSEQUENCES ARE INEFFECTIVE

- Persons with PWS are predisposed to react stubbornly to negative consequences.
- Persons with PWS can shut down completely in response to a negative consequence.
- Punishment does not change future behavior.
- A person with PWS often has the ability to adjust to the most unpleasant situations. Some caregivers perceive this as the need to apply more and more severe consequences.
- Some people with PWS like to “stir the pot” and will try to make you apply a negative consequence just to see what happens.

PUNISHMENT



- Punishment does not teach a new skill. Punishment does not serve to teach someone with PWS but instead escalates unwanted behaviors and creates a meltdown.
- An overall lack of impulse control does not allow the person with PWS to see the whole sequence of events.
- An inability to change behavior based on a negative outcome (i.e. natural or applied consequence) in past similar experience makes it less likely to learn from prior situations.
- Never use corporal punishment – never physically hit.
- **Never use food as a punishment.** Food should be as stable as oxygen.

SO WHAT DOES WORK?

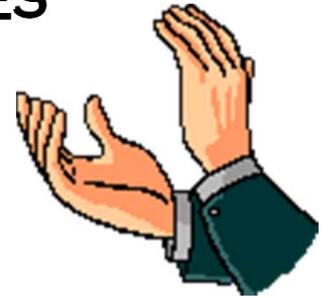
“The most effective form of consequence management is one in which the desired behavior must be earned rather [than] undesired behaviors punished. For example, four hours without a tantrum may earn 25 points and 100 points are necessary for a phone call.”

~Barbara Whitman,
“Understanding and Managing the Behavioral and Psychological Components of Prader-Willi Syndrome”

A positive behavioral strategies are the most effective
for persons with Prader-Willi syndrome

REWARDS, INCENTIVES, POSITIVE BEHAVIOR STRATEGIES

- Praise! Praise! Praise! to motivate desired behavior and teach new skills.
- Use positive reinforcers and incentives: stickers, small prizes, special time, being a special helper, etc.
- Keep the individual with PWS thinking, not going into feeling mode.
- Avoid power struggles because you won't win!
- Clearly and calmly ask for what you want in a way that is understood by the listener (don't leave it at what you *don't* want).
- Make your request in such a way that there's no "bad" choice.
- Don't give in! Giving in today over a "little thing" will lead to a large tantrum tomorrow. The peacefulness and overall quality of life of everyone depends upon upholding the established limits. Think long term!



WHAT ELSE WORKS?



- Disguise “No” as a positive. Spin it!
 - Child: “I want to play outside”
 - Parent: “Me too! Looks fun out there! After homework’s all finished, let’s go outside!”
- Logic and reason will generally not prevail when the person with PWS gets upset and/or “stuck” on an idea or position. Show empathy by repeating back what you’ve heard he feels/wants to help get “unstuck.”
- In times of distress, use the broken record technique. Use empathy. Be gently and compassionately unswerving.

AND...

- Be smart. Think two steps ahead of the individual with PWS. Anticipate what may cause anxiety and work to eliminate or reduce the potential stressors.
- Be a Mamma Duck... Lead, don't wait for person to initiate movement.
- Use music and fun 'games' to hurry along/motivate desired behavior.
- Keep the person with PWS busy! Downtime is fine; boredom fuels problems.
- Distraction can be very helpful.



AND...

- Don't promise anything that you cannot or will not do.
- Genuine Time Out's can help the person gain control over his body and help reduce an escalation or "drama" of a situation.
- Use CPS and cooperatively-created contracts.
- Use empathy. Be gently and compassionately unswerving.
- Create consequences that are more "in the moment," "natural."
- In times of distress, use the repeat method and/or broken record technique.
- **Have appropriate expectations.**

AND ...

- Never use food as a reward. Never use food as an incentive. It is ok to schedule a meal or snack or a *scheduled* treat after a task.
- Ignore inappropriate behavior. Reward appropriate behavior.
- Schedule non-preferred tasks/activities before preferred tasks/activities.
- Listen to the person who needs to talk. The time it takes to listen and express *genuine* empathy is worth it. Repeating the concern you've heard is often enough to reduce the hyper-emotion attached to it.
- **Be patient! Be patient! Be patient!**

SIDESTEPPING BEHAVIOR PROBLEMS



- Building up to an argument? Don't engage! If the person with PWS is building up to a disagreement, calmly state your desire not to talk about it anymore. Or guide him to a private area and discuss it there. Allow the individual to have the last word. Try to make the statement, "I will make one more comment and then you can make one more comment and then let's talk about something else."
- Once a meltdown has begun there's nothing to do but wait it out and work to keep the individual safe. Persons with PWS generally feel very remorseful and ashamed of their behavior after a meltdown. Comfort the individual without condoning the unwanted behavior.
- Later, when everyone is calm, it may be possible to go back and examine what happened in order to learn how to avoid the behavior meltdown next time.

MELTDOWN REMORSE

- If a meltdown begins, there's nothing to do but wait it out and work to keep the individual and others safe.
- After a meltdown or behavior outburst people with PWS often feel sorry, embarrassed or shameful. Comfort the individual without condoning the unwanted behavior.
- Accept an apology if offered; don't let the absence of an apology become yet another power struggle.
- The intensity of the person's remorse does not reduce the likelihood of the same thing happening again.

DON'T FORGET...

- Be smart. Think two steps ahead of the individual with PWS. Anticipate what may cause anxiety and work to eliminate or reduce the potential stressors.
- Assistance is generally needed to stay on task, understand directions, transition from task to task, manage potential frustrators.
- Multiple transition cues are often necessary to move from one activity to the next.
- Persons with PWS learn best by repetition. Directions and tasks may need to be broken down into smaller steps and taught a few times .
- Always look and act calm on the outside no matter what you feel on the inside.



EXCESSIVE TALKING? REPEATED QUESTIONS?

Determine the function of the excessive talking or repeated questions.

Are repeated questions a function of a short term memory deficit?

Answer the question 2-3 times then ask the individual to tell *you* the answer to their question.

Is the function to reduce anxiety?

If Yes, then strategize how to reduce the anxiety.

- What is contributing to the anxiety?
- Unknown upcoming event or situation? Paint a picture of what to expect.

EXCESSIVE TALKING, REPEATED QUESTIONS CONTINUED

Is the function a reflection of a learning style or a learning disorder (i.e., nonverbal learning disorder)?

If Yes, then know the individual may need to talk out loud in order to better understand their situation or environment. Gently guide their understanding with reflections, clarifying questions or statements.

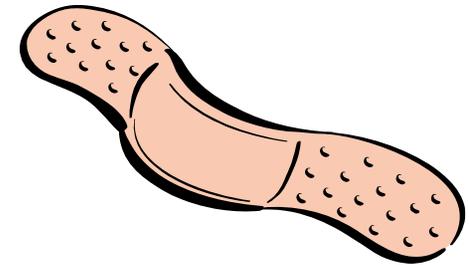
Is the function to engage in social interaction?

If Yes, then strategize how to improve social interaction/social skills:

- Engage the individual in conversation, steer into a dialogue (from a monologue), move from topic to topic.
- Help individual engage with/play with peers.
- If you need a break from listening, politely tell the person you need a break from listening and ask them to stop talking for specified period of time (i.e., 5 minutes)

SKIN PICKING

- Skin picking is a biochemical drive that occurs in >90% of persons with PWS
- Keep nails short
- Keep pick area covered with band-aid if possible
- Keep pick area slick with lotion
- Keep hands busy! Squishy balls, toys, coloring, hand grips, crafts, etc.
- Use bug spray and cover bug bites and scabs with a band-aid straight away
- Use the rolling technique as described by BJ Goff, PhD
- Some medications e.g., Topiramate (Topamax) can be helpful with severe picking cases



PSYCHOTROPIC MEDICATION SHOULD BE CONSIDERED...

When all of the environmental factors that may be causing anxiety and stress are managed

AND

When the unwanted behaviors continue

AND

When the unwanted behaviors significantly interfere with the quality of life of the person with PWS ...and the family

When all of the above exists, then it's time to consult a psychiatric physician.

Provide the M.D. with information about PWS available from PWCF.



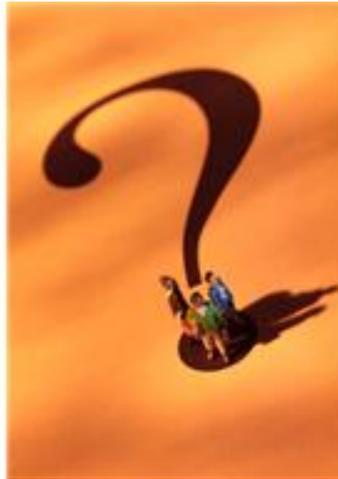
SOCIAL SKILLS

- Almost all persons with PWS are highly social and crave emotional connection.
- Most people with PWS are highly egocentric which interferes with social skills acquisition and the qualities necessary to develop and maintain peer relationships.
- Most children with PWS are drawn to adult relationships because adults are typically easier to connect with than same-age peers.
- Repeated questions, excessive talking may be ways to initiate social interaction. *Encourage dialogue, not monologue.*



It is essential to specifically teach social skills and provide opportunities for practice.

QUESTION & ANSWER EXCHANGE





Prader-Willi California Foundation

Supporting People with Prader-Willi Syndrome

Prader-Willi California Foundation
514 N. Prospect Avenue
Suite 110-Lower Level
Redondo Beach, CA 90277
310-372-5053 • 800-400-9994
Fax 310-372-4329
info@pwcf.org
PWCF.org



