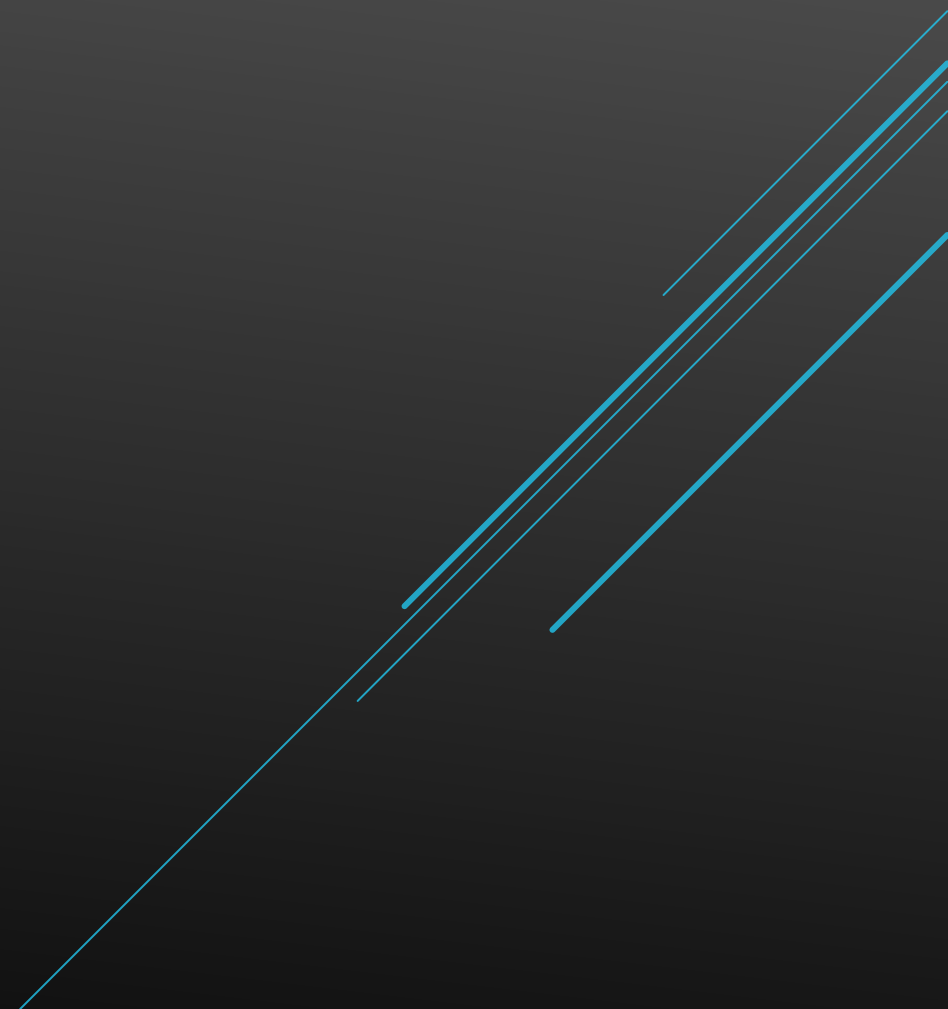


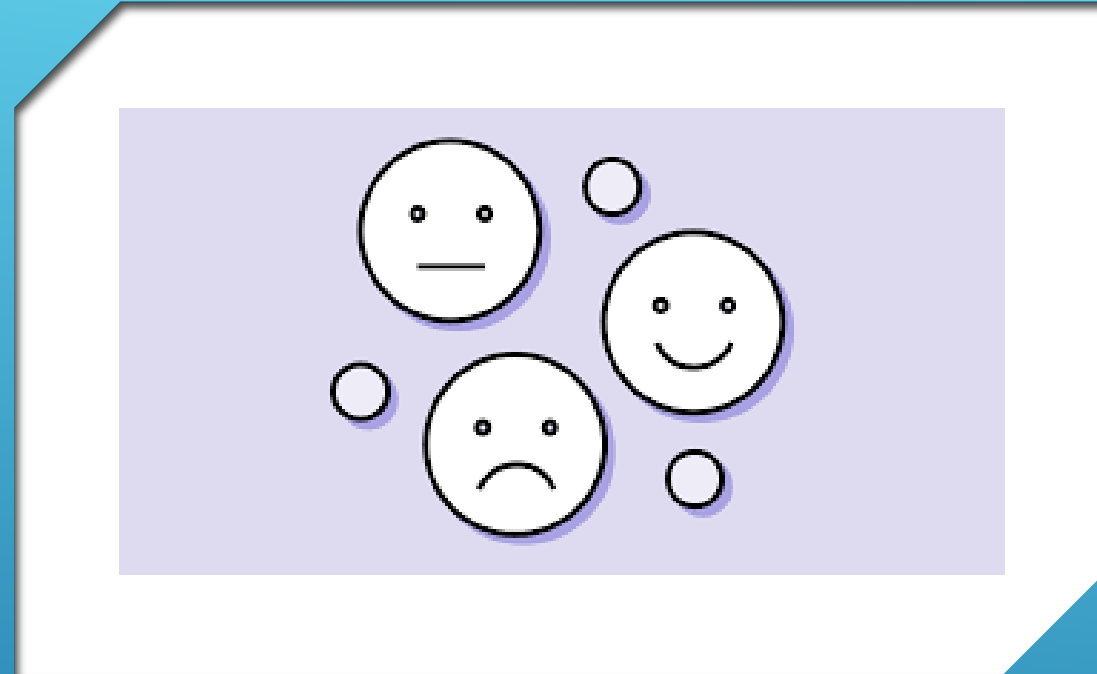


# SUPPORTS FOR SUCCESS

For People With Prader-Willi Syndrome

# OUTLINE

1. Behavioural presentations of PWS
  2. No Hope, No Chance= No Disappointment
  3. Environmental Safeguards
  4. Support/Management Strategies
  5. Resources
- 



# BEHAVIOURAL PRESENTATIONS

Strengths & Challenges

PWS is a spectrum disorder, so not all of the behavioural characteristics discussed will apply to the person you know. There will be differences between others with PWS as everyone is unique in their own way. These are more general behavioural and cognitive presentations of PWS that help guide the best supportive environment.



# COGNITIVE STRENGTHS

- ▶ Long term memory
- ▶ Receptive language
- ▶ Ability to learn from visual materials
- ▶ Reading
- ▶ Hands on learning

Chedd, N., Levine, K., & Wharton, R.H. (2006). Educational considerations for children with Prader-Willi Syndrome. In M.G. Butler, P.D.K. Lee & B.Y. Whitman (Eds), *Management of Prader-Willi Syndrome* (3<sup>rd</sup> ed., pp.302-316)



breakfast



arts & crafts



play time



Evening

**PM**



lunch





# COGNITIVE CHALLENGES



- ▶ Short term auditory memory
- ▶ Fine motor skills (pulling a zipper, tying shoes, holding a pencil, etc. )
- ▶ Expressive language (telling us their thoughts, feelings using words, gestures, writing, etc. )
- ▶ Abstract thinking (time, considering someone else's point of view, "don'ts" jokes, teasing, etc.)
- ▶ Auditory processing (learning through verbal instruction is difficult for many; they may take longer to respond in conversations, and have trouble following verbal directions)

# THINKING PATTERNS

- ▶ Rigid thought processes
  - ▶ Receive and store information in an orderly manner.
- ▶ Perseverative or obsessive thinking
  - ▶ Tendency to get stuck on one topic or thought, often accompanied by anxiety.

**"Success doesn't  
come from what  
you do  
occasionally. It  
comes from  
what you do  
consistently"**

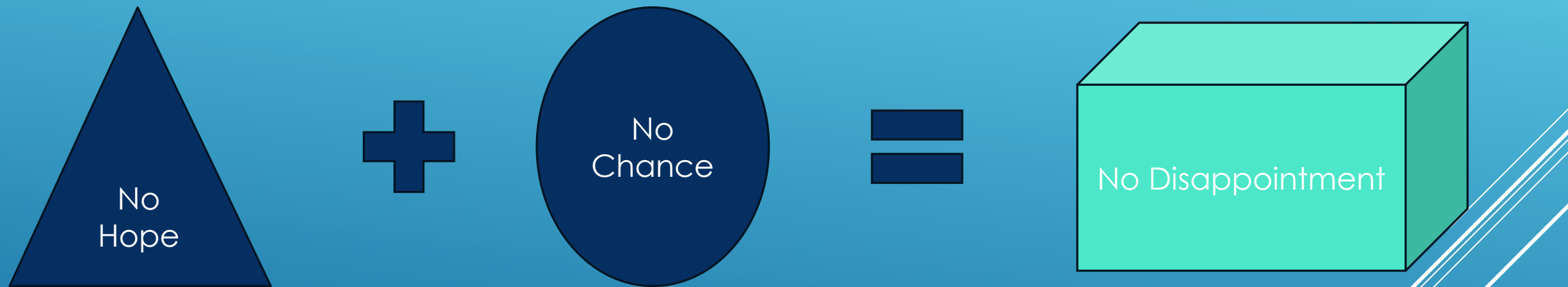
MARIE FORLEO

# EMOTIONAL/MENTAL HEALTH





This principle forms the foundation of effective support strategies for people with PWS. It is often related to food security, but it applies to all aspects of their lives. We need to be very clear, specific, and concise when providing directions, information, or answering questions.



There should be no hope of accessing something different from what was planned or answered (this could be food, answers, information) and no chance or opportunity of accessing other than what was planned which results in no disappointment related to false expectations by the person with PWS.

► One of the major way to apply this principle is through environmental supports. This refers to both the physical environment as well as interactions with others.



## ENVIRONMENTAL SUPPORTS



NO FOOD  
ACCESS

Several thin, white, parallel diagonal lines are drawn across the bottom right corner of the blue background, extending from the right edge towards the center.

► With the principle of No Hope No Chance No Disappointment in mind, have all food locked to prevent any chance or hope of accessing it.

This may require limited access to certain areas of the person's home, like the kitchen, pantry, and garage. If there is any chance of accessing food the person will only focus on accessing that food and creating unnecessary anxiety.

► This includes no access to credit/debit cards, cash, online orders, etc.





NO FOOD  
STIMULI

The presence of food stimuli often provides a sense of anticipation or hope that food may soon be available. Which often leads to disappointment. This includes not talking about food. This can be difficult because food is at the center of many of our social gatherings. We need shift gears and focus on other aspects of holidays and social gatherings.

For example, birthdays can focus on non- edible gifts, fun activities, reading cards (with no food stimuli) given to them, etc. For Christmas, focus more on decorating the tree, walking around the neighbourhood to look at the different decorations, similarly with Halloween, focus on costumes, decorating pumpkins if tolerated, etc.

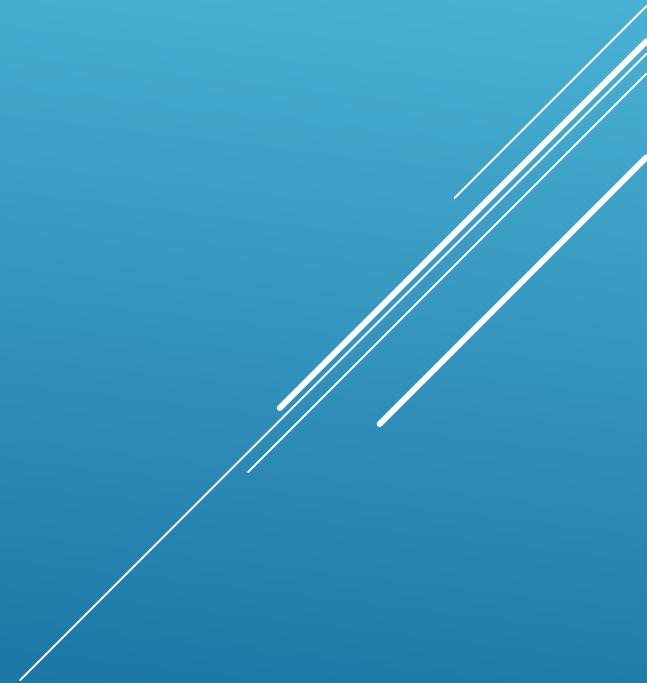




SUPERVISION

Supervision is imperative especially in an environment that you have little control over, i.e. Relatives' homes, hospitals, medical offices, etc.

Supervision helps create a secure environment for the person, reducing anxiety by removing the hope or chance of accessing extra food or items., or different expectations, thereby preventing potential disappointment.





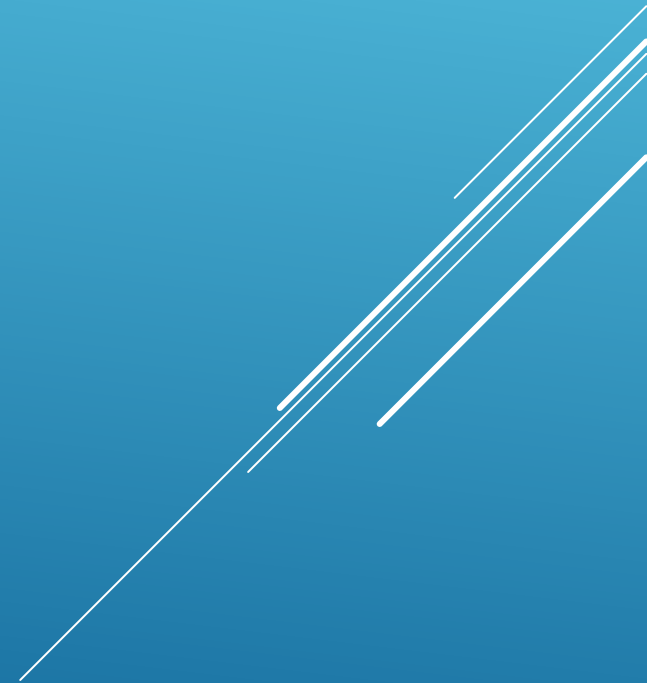
SEE THE  
ENVIRONMENT  
WITH  
DIFFERENT EYES

To support the principle of “no hope, no chance, no disappointment” it is essential to consider the environment from the perspective of a person with PWS.

This approach requires proactive planning and thorough scanning of the surroundings or conceal any food related items that may trigger anticipation or anxiety.

For example, for medical visits you would want to look at maps to see what the best entrance to use would be, where there are minimal food stimuli. If possible, visit the place to do the scan.

Environments such as buffets, restaurants, and grocery stores are typically not advisable, as they can be overwhelmingly anxiety-provoking. It would be unfair to place someone with PWS in a setting that elicits such high levels of stress.





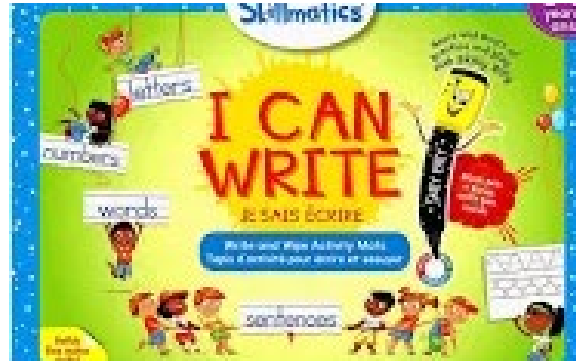
# SUPPORT STRATEGIES



**CONSISTENCY**  
is a key

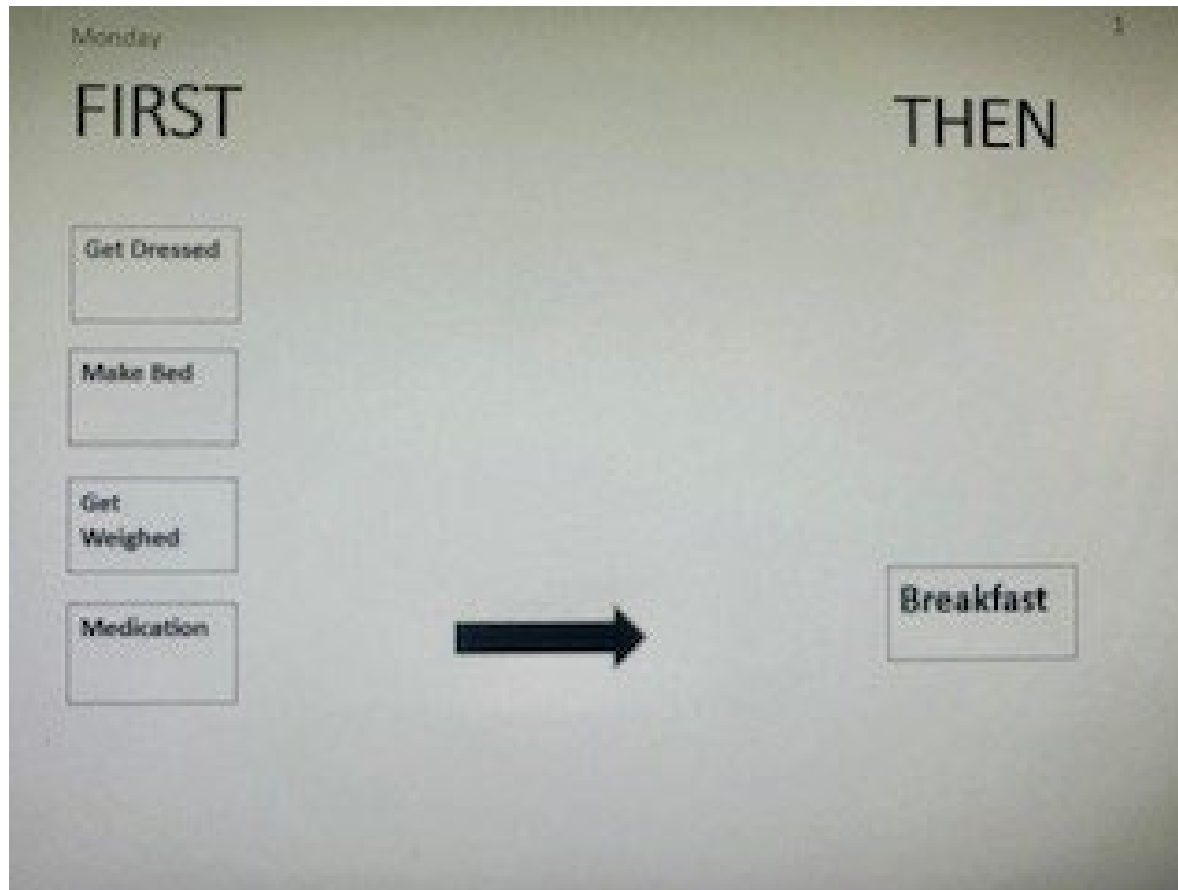
- ▶ Consistency and routine leads to predictability which is key to reducing the anxiety a person with PWS experiences. When they know what to expect they feel less anxious.
- ▶ This can be accomplished by the creation and consistent follow through of rules, and schedules which provide clear expectations.





# DAILY ACTIVITY SCHEDULE (DAS)

- Consistency and routine leads to predictability which is key to reducing the anxiety a person with PWS experiences. When they know what to expect they feel less anxious.
- This can be accomplished by the creation and consistent follow through of rules, and schedules which provide clear expectations.



# FIRST...THEN

►To help keep the person motivated it is best to structure the schedule using the first....then procedure.

►First identify the preferred activities that the person looks forward to and then identify the non-preferred activities that the person needs to do but does not particularly like doing... we all have those. So basically, the person completes some non -preferred activities before the preferred activity. This will give them something to look forward to after doing some of the non- preferred tasks.



















►It's also important to make the non-preferred activities as interesting or fun as possible.

►Dependent on the person's tolerance, some schedules can include times while for others it increases their anxiety, so their schedules are more activity based.

# WHY IS THE DAILY ACTIVITY SCHEDULE SO IMPORTANT?

- ▶ Keeps the person busy, thereby reducing opportunities for perseveration
- ▶ Provides predictability
- ▶ Provides opportunities for learning new skills



 Get dressed		 Get weighed		 Make Bed	
 Room Time		 Breakfast		 Brush teeth & swab	
 Circuit Training		 Snack		 Baseball	 Sticker!

►To address the difficulty with auditory processing the use of a visual schedule is recommended. This example uses pictures with labels. Dependent on the person's reading ability the schedule can be just pictures or text.

►The visual activity schedule has many teaching functions, such as learning to read, follow a sequence of activities, and plan.

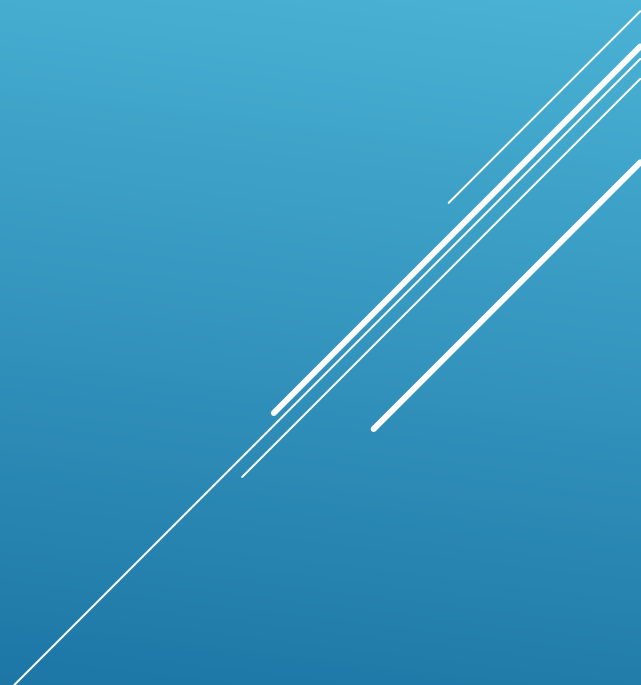
## VISUAL ACTIVITY SCHEDULE



# TRANSITIONS

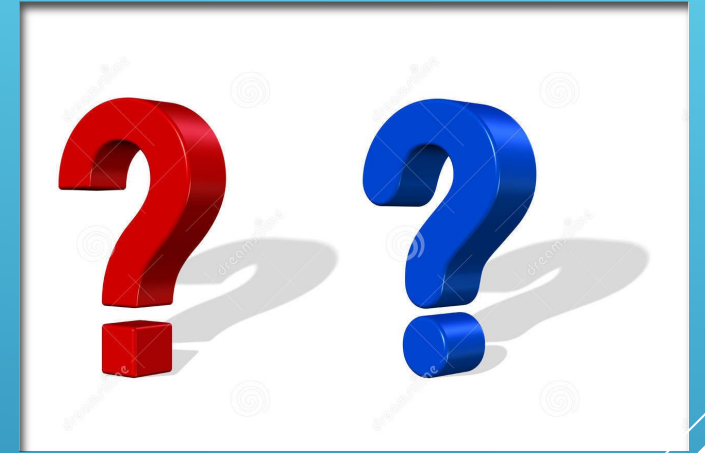
- ▶ Visual Timers & Prompts
- ▶ Visual Cues

- Transitions are difficult for most people with PWS, especially the transition from a preferred activity to a non preferred activity.
- Transitional prompts are very helpful by providing a prompt at 5 minutes and then at 2 minutes to let them know that the end of the activity is approaching. Use of visual timers and cues need to supplement the verbal prompt 5-minute and 2-minute prompts.
- This “Time timer” clock is helpful with the abstract concept of time, as it shows time decreasing in a concrete way.





- ▶ Repetitive questioning or perseveration is another common characteristic of PWS, which we learned can result from heightened anxiety. Repeatedly responding to the same questions is not helpful and tends to increase their anxiety.
- ▶ Keep in mind the “No hope, no chance, no disappointment” principle, when answering any questions to help reduce repetitive questioning from happening.
- ▶ Provide a clear answer. If it is no, say “No”. Ambiguity leads to more anxiety which can lead to escalated behaviours.



## RESPONSE TO REPETITIVE QUESTIONING

### Person Asks the Question A Second Time

Minimize Distractions

Let the person know that this will be the last time you will answer the question

### Person Repeats the Question

Neutrally remind them that you answered that question.

Do they remember the answer?

Then we are done talking about it.

### Person Continues to Repeat the Question

Do not respond to the question

Focus on activity or task at hand

## 2 QUESTION RULE



► Skin picking is common in people with PWS, approximately 81% of adults with PWS. (Management of Prader Willi Syndrome, 3<sup>rd</sup> Edition, 2006). This behaviour tends to increase in times of increased anxiety, so again providing predictability help alleviate the additional anxiety.

► Keeping their hands busy, moisturized and not providing attention to the picking behaviour can decrease the intensity and frequency. Skin picking is common in people with PWS, approximately 81% of adults with PWS. (Management of Prader Willi Syndrome, 3<sup>rd</sup> Edition, 2006). This behaviour tends to increase in times of increased anxiety, so again providing predictability help alleviate the additional anxiety.

## SKIN PICKING



- ▶ Excessive acquisitions and use of items is common in PWS and likely due to the lack of impulse control and never feeling satiated.
- ▶ External measures such as limited access helps with the lack of control.
- ▶ For example, limit access to toiletries by keeping them out of sight and only available when needed.
- ▶ Provide measured/reasonable amounts of items. i.e., one puzzle or game at a time, measured shampoo and conditioner, etc.
- ▶ Hoarding behaviours are compulsive symptoms that are found in about 60% of the PWS community (Management of Prader Willi Syndrome, 3<sup>rd</sup> Edition, 2006) .

## EXCESSIVE ACQUISITIONS & USE

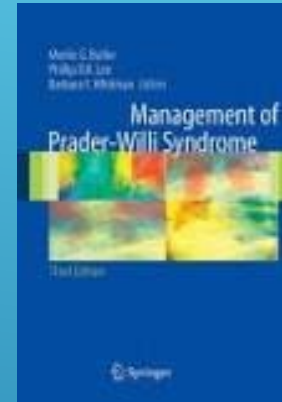
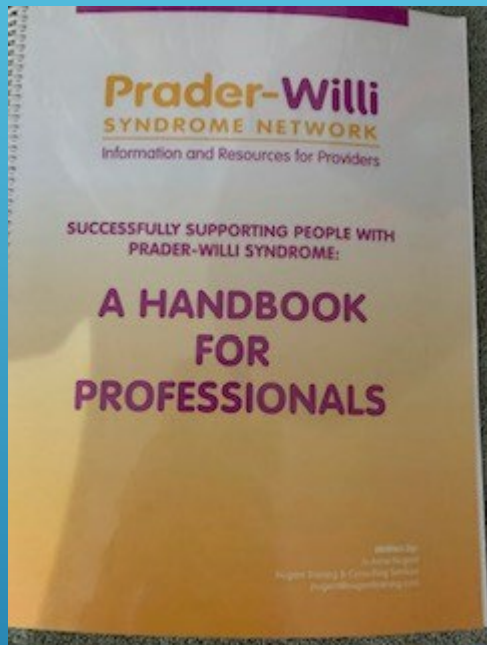


# POSITIVE REINFORCEMENT

- Positive reinforcement is very effective in increasing the behaviours we want to see more of .
- You want to catch them behaving well and provide praise and or a reward, no matter how small.
- The key is to find something that the person enjoys enough to motivate them and then providing that when they engage in the appropriate behaviour.
- Depending on the person you can use an activity that the person really enjoys as a reward.
- Others enjoy puzzles, watching music videos, or going to the movies.
- You can use token systems whereby the person earn points for socially appropriate behaviours like following instructions, staying calm and focused, using their calming tools when feeling angry or anxious. When they achieve a specific number of points, they earn the reward they chose.
- These rewards may change and that's good, but we have noticed that they tend to stick to what they know.







# RESOURCES

