

The Behaviour Phenotype in Prader-Willi Syndrome



“Is this behaviour
part of the
syndrome”?

Most people with Prader-Willi Syndrome will exhibit a set of behaviours which are so characteristic of the syndrome that they are known as a behavioural phenotype. Most are linked to the hypothalamic dysfunction and/or cognitive problems. However, in terms of severity, these behaviours vary considerably from person to person, over time and depending on circumstances.

Until recently, we referred to ‘Challenging Behaviours’, however we now know that this can put a negative label onto the person displaying these behaviours PWSA now refer to ‘Behaviours of Concern’.

In order to understand the behaviours we are discussing and the Positive Behaviour Support (PBS) approach first developed in the 1980’s, we need to look back to the introduction of the PBS approach in care settings in 2014 through the Department of Health guidance - Positive and Proactive Care. This document proposes that care organisations use a PBS approach to support those with learning difficulties who display behaviours of concern. This guidance was produced following a review into the Winterbourne View scandal.

Challenging behaviour was defined by Emerson (1995) as “culturally abnormal behaviour(s) of such intensity, frequency, or duration that the physical safety of the person or others is likely to be placed in serious jeopardy. Or behaviour that leads to a poorer quality of life by impacting their ability to join in everyday activities.

Evidence has shown that PBS-based approaches can enhance quality of life and also reduce behaviours of concern. It’s about identifying triggers for behaviours; managing the environment to reduce triggers and implementing strategies to help people develop the skills they need to reduce behaviours.

This document explores the behavioural phenotype, in PWS, and suggests strategies which may help you to manage them. However, your in-depth knowledge of your son, daughter or individual you care for, will support your decisions as to how and when to apply different types of management techniques.

It is important to remember that we are focusing on behavioural concerns in PWS, people with the syndrome have many positive characteristics which often offset their difficulties to a large extent. As you know they are friendly, sociable, kind and caring, and many have a wonderful sense of humor. Each individual will also have other traits which are equally positive. Good behavioural management can bring these positive traits to the fore even more.

Remember:

All behaviours happen for a reason and are usually a response to an unmet need
These behaviours are not learned and are often caused by immature emotional and social development

Typical behaviours in PWS - the behavioural phenotype

Behaviours in PWS have been researched so much that they have become a phenotype group of symptoms which apply specifically to PWS. Many are directly related to the main characteristics of the syndrome such as:

Hyperphagia - the excessive and overwhelming appetite

Hypogonadism - the immature sexual development

The individual with PWS may exhibit some of these behaviours and below we show the typical behaviour; what that might look like and suggest strategies to manage it:

The behavioural phenotype

A description of the behaviour

Suggested strategies

Food seeking/stealing

Hyperphagia typically begins from age 2-5. It can start as an interest in food, but can develop into a constant desire for food leading to this behaviour. Brain scans show us that the signal to the brain, which indicates that we are full, is not present in individuals with PWS.

- Seeking out food in any environment
- Negotiating food from others
- Helping themselves in public spaces

- Remove all opportunities to obtain food
- 1-2-1 supervision wherever and whenever possible

“Lying”, blame shifting and story telling

Often known as ‘confabulation’, children with PWS will often have their own version of events which they believe to be true and alongside this, they will tell very convincing stories.

However, if they have been food seeking or stealing, they will often deny any involvement, even when confronted with the evidence.

- Having their own version of events
- Denying involvement against any evidence

- It is important to distinguish between harmless imaginative stories and what is real.
- Let them know that you know the truth

Skin Picking

70% of people with PWS do this, it is a coping mechanism. (We all have a tendency to do this, fiddle with our hair, pick a scab when it is about to come off but we know when to stop, a person with PWS doesn't). It is thought that they find skin picking pleasurable, it releases endorphins into their systems. Possible causes: boredom, anxiety, attention seeking.

- They may have a small spot or bite or graze and they pick at it.
- Skin picking can be more serious if it becomes internal picking

- Cover affected area, keep nails short, use emollients e.g. Vaseline
- Massage skin with soft brush
- Redirection techniques - give something to do with hands - a fiddle toy, bubble wrap.
- If it is nail biting, paint the nails and make them pretty
- Take photos of improvements and use lots of praise

▪ Outburst of rage or prolonged meltdowns

This behaviour can be linked to learning disabilities and immature emotional and social development in individuals with PWS. It is usually a way for them to communicate an un-met need or a response to something that they find difficult to cope with such as change, anxiety or frustration.

- Shouting, crying, screaming.
- Physical and verbal aggression.

- Remain calm and avoid raising your voice or showing anger.
- Ensure the safety of the person, others and yourself.
- Show understanding e.g "I can see you're angry".

Stubbornness/difficulty in adapting to changes in routine

Individuals with PWS can appear to be very stubborn, this is often because they have not been allowed the time they need to adapt to changes in routine and to process the information. Or it may be that their expectations have not been met.

- Refusal to cooperate.
- Frustration and anger or becoming withdrawn, avoiding eye contact.

- Need to know what is happening and when, preferably in advance of changes.
- Give instructions in a variety of ways. Allow them time to process the information.
- If you want a child with PWS to move from one activity to another, prepare with "Now and Next" and/or visual

Argumentative or oppositional behaviour.

Again, this behaviour can be triggered by their dislike of change and situations that are not what they had expected and wanting things 'their way'. For example, if swimming is Tuesday at 2pm then it is always Tuesday at 2pm. When things are not what is expected, they can get stuck and find it difficult to shift their focus.

- The need to say NO before they will say YES!
- Wanting things their way every time.

- Allow some level of choice and control.
- Let them make decisions between two options rather than unlimited choice which can be overwhelming.
- Prepare them for change using visual countdowns such as sand filled egg timers.

Obsessive and/or compulsive behaviour - keeping on with the same topic (perseveration)

This behaviour is very common in PWS and you are probably very familiar with it. Often in regard to food, but it can also be other topics or people. However, it can be difficult to deal with as the person needs to be reassured that you have listened and understood

- Asking the same question over and over again, sometimes with subtle differences.
- Talking about something incessantly.

- Ask them to repeat back the answer you have given.
- Reassure them that you have heard and that you understand.
- Try saying - "You can ask me/tell me three more times", and your fingers to count down as a visual reminder.

Hoarding and collecting

Lots of people with PWS like to collect things to keep and this can also fall into the obsessive behaviour category. They may place a great deal of importance of these things which can make getting rid of something very difficult.

- Collecting books, magazines, jigsaws, certain toys etc.

Negotiating can work well:

- One item in and one out, (donate to a charity shop).
- Allow a set amount of space in a drawer or set aside one particular shelf.