

## Challenging behaviour in PWS

Children and adults with PWS are usually happy, friendly, sociable and loving individuals, so it can sometimes come as a shock if they exhibit violent or very aggressive behaviour, or when emotional outbursts (sometimes called melt-downs) begin to escalate into physical aggression.

We do not have any definite figures to say how common challenging behaviour is in PWS, especially as it can occur in phases, affecting individuals at different times and in different circumstances. We do know that it can occur in very young children and at any age thereafter.

## Understanding challenging behaviour in PWS

Challenging behaviour in any one, regardless of whether or not they have PWS, usually results when someone has reached the limits of their endurance of a situation. How much any one person can stand varies considerably between all individuals.

People with PWS are much more vulnerable to stress than the general population. Generally speaking, they have a low tolerance for anxiety and frustration. They also have poor control over their emotions, acting impulsively. Most find it difficult to deal with change. They have difficulties processing information, so that it takes longer for them to work out what is happening and why. The combination of these factors means that acting out is more likely to happen in PWS, and sometimes this results in aggression.

However, the onset of mental health problems such as psychoses and an escalation of anxiety and mood disorders can also be exacerbated by stress, and consideration must be given to this as a possibility, if the behaviour cannot easily be attributed to other triggers.

Certain medications may also give rise to more challenging behaviour. Testosterone as a hormone treatment for males is sometimes mentioned anecdotally as a cause, but research has so far not discovered a distinct relationship. There are case reports suggesting that oestrogen therapy for females can increase mood lability as well. The use of SSRI antidepressants and atypical neuroleptics has been linked with mood and behavioural activation that can present with impulsive behaviour, aggression, self-injury, and property destruction.

## Triggers

These are some of the more general triggers, not only for challenging behaviour, but for emotional outbursts of any kind:

- Anxieties around food; presence of food in the room or nearby
- Frustration at not being able to do what he or she wants to do

- Communication or speech and language problems
- Feeling under pressure (whether or not the pressure is actually there – eg, saying “she forced me to do it” when in actual fact the person was merely making a suggestion about what they the person with PWS might like to do).
- A “chaotic” environment with too much stimulus and/or too little structure
- In a low or unstable mood
- Unrealistic expectations on the part of others
- Real or perceived unfairness in a situation
- Change of any kind
- False expectations or disappointment
- Bullying, or physical or sexual abuse by others
- Harsh tone and punitive attitudes from carers
- Additional undiagnosed medical and learning disability conditions (eg autistic spectrum disorder)

However, there are many other individual triggers. Finding out what is the cause can be difficult, because of the propensity of people with PWS to shift blame or give another reason for an outburst than what was the real cause.

## Preventing or minimising aggressive and violent behaviour

### Create the right environment

In view of all the above considerations, the environment for someone with PWS has to be “right”- but it is up to others to provide the environment in which they can thrive. It is most likely the environment, or other people in that environment, which are the cause of challenging behaviour when it only occurs in certain situations, eg at school, but not at home, or vice versa.

For many individuals with PWS, their behaviour will stabilize when in a structured, low-stress environment, where food access is controlled, boundaries are clear and a consistent approach is adopted by everyone involved.

Everyone working with the individual should be aware of potential triggers and how to avoid them.

### Recognise signs of emotional distress and act before the situation escalates

Each individual may have their own way of showing that they are becoming distressed: rubbing or shutting eyes, wringing or waving hands etc. Over time you will recognise what they are, and sometimes it is possible to take the person to a calm, safe space, acknowledge that you have recognised they are becoming upset, and address any issues they may have – particular if these are relatively minor.

## Teach coping skills and behavioural relaxation techniques

### Coping skills

Coping skills should be embedded into the daily programme of activities for people with PWS. They should also be scripted and rehearsed with carers in residential care and other situations where the person is away from home. Then, when the carer perceives an escalation, they can cue the person to use one of

the practiced techniques. In this way, they can draw on these before they begin to go into meltdown. These skills include:

- Deep breathing
- Stress relief through sensory balls
- Listening to music
- Learning how to communicate feelings appropriately
- Taking a break

### **Behaviour relaxation techniques**

Learning simple breathing control exercises and how to tense and release different muscles groups throughout the body can help reduce feelings of anxiety arousal or calm an individual who is feeling annoyed. Again, these techniques must be scripted, rehearsed, and cued by carers and imbedded in the daily program.

**Adopting a Relaxed Posture** - Encourage the person with PWS to sit comfortably in an arm chair, with arms should be placed palms down along the arms of the chair in a resting position. Encourage the person to rest their head back in the chair and put both feet up on a foot rest, and take slow deep breaths and close their eyes. (You may need to demonstrate how to do this and praise when it is done correctly)

**Tensing and Releasing** – Show the individual how to tense the following muscle groups in turn. Each tensed position should be held for a slow count of 10, and then relaxed so muscles go floppy like jelly. Repeat each position at least three times.

- Hold arms out and clench fists
- Pull shoulders up to ears
- Push both arms down on arm rests
- Strongman (tense biceps)
- Push both knees together
- Push head back hard into head rest
- Open mouth and eyes wide
- Close eyes and scrunch up face

**Abdominal Breathing** – Encourage the person with PWS to lay back in a comfortable chair with their feet up, hands on belly, with fingers interlinked. Show them how to breathe deeply into their belly, not their chest, so that their belly moves in and out. You'll know if they are doing this correctly as their interlinked fingers will separate slightly.

**Breathing Elephants** – Ask the person to take a very deep breath in and as they exhale, try and count as many elephants as they can until they run out of puff “one elephant, two elephants, three elephants...”etc. (They should not continue to breathe normally as he counts, the object of the exercise is to regulate breathing). This is a good, fun exercise which can be done as a competition with another person to see who can count most elephants before they run out of breath!

## Managing challenging behaviour

The person who is being attacked is not necessarily the cause of the aggression, which is often focussed onto the nearest available object or person.

Your immediate response and throughout should be calm and low key, ignoring screaming and shouting. Do not do or say anything which might make the situation worse. Be aware that people with PWS are very sensitive to tone of voice; even the slightest hint of irritation, cajoling, or frustration on your part will be picked up by them. Do not argue with a PWS person; this will only escalate or prolong the incident. Threats and bribery are ineffective.

It is futile to attempt to reason with a PWS person in the middle of an outburst, e.g., saying “things aren’t that bad” will not work - the thinking and problem solving part of their brain is “offline” at this time.

Make the environment as safe as you can, and direct other people away from the incident.

### After an incident

The person with PWS will usually need time to recover their equilibrium. So provide a calm, low stimulating space, where they can go and lie or sit quietly. Many often go to sleep.

Wait until everything is calm again, and then, if possible, try to address the issue in a sensitive manner and find out what caused it. This is not always possible, however, because the individual may become upset again, feel under pressure or be unable to identify a cause. You may have to wait until the next day for the person to be calm enough to respond without the situation escalating again. What may seem a very minor issue can make a person with PWS very anxious or frustrated, so sometimes it is relatively easy to ensure that issue is addressed, but often there are more deep-seated concerns.

The most important thing after an outburst resolves is to get the person back into the program of the daily schedule as soon as possible.

## Parents - Help for you

If your son or daughter is displaying challenging behaviour it can be very stressful. He or she may direct this behaviour towards yourself or other family members, or it may be directed at children and teachers at school or college, or staff in day centres or residential homes and supported living. Many describe their situation as “walking on eggshells” - never sure when the next meltdown will occur, or, if the phone rings, if it will be yet another request to collect your child from school or college, with a threat of, or actual, exclusion.

The important thing to remember is that PWS is a very complex disorder and that your parenting skills are not necessarily at fault. You may need help from experts (see below) who can carry out a thorough analysis of what is happening with your child and recommend management strategies or treatments.

On very rare occasions, this may mean moving the child or adult out of their present circumstances into something more appropriate (eg from mainstream to special school, or from supported living to residential care), or into a specialist treatment centres. Again, this is not an indication of failure on your part, and may

well be in your son or daughter's best interests, providing them with an environment in which they are better able to cope.

Recognise when you are reaching a situation in which you personally are unable to cope:

- Feeling helpless or powerless
- Breaking down in tears
- Dreading phone calls
- Feeling frightened of the person with PWS
- Wanting to run away

Talk to your GP or call social services and ask for help. Remember too that the PWSA UK is here to offer a listening ear, or to support your requests for help from the authorities. You can reach us by phone on 01332 365676 (Mon – Fri, 9 am – 5 pm, answerphone at other times), email [admin@pwsa.co.uk](mailto:admin@pwsa.co.uk) or on Facebook (search for Prader-Willi Syndrome Association UK).

## Help from experts

If you have tried everything, or the aggression is becoming a danger to others or the person themselves, ask your GP, community nurse, consultant or social services for a referral to psychiatric services or the CAMHS (Child and Adolescent Mental Health Services) in your area. When you go for the referral, make sure you take along information about PWS, including this leaflet, and this very helpful article for psychiatrists written by Janice L Forster and Linda M Gourash from the USA

<http://www.pwcf.org/wp-content/uploads/2012/02/Psychiatry-Primer-for-PWS1.pdf>

If the psychiatrist has little or no experience in working with someone with PWS, we can provide contact details of someone they can speak to for more information.

In some cases, medication may be prescribed. As a general rule, this should be at a lower dose than normal.

© PWSA UK