

Behaviour management Adults



Introduction

Almost everyone with Prader-Willi syndrome presents with behaviour which can bring challenges to either themselves, or those around them, or both. However, this varies considerably between individuals.

Behavioural challenges range from very mild to very severe, and each individual may display differing behaviours. These

in turn may vary with age, external environment and emotional development.

The degree of learning disability does not necessarily affect the severity of behavioural challenges. Those who have only mild learning disabilities are just as likely to have challenging behaviour as those with a severe learning disability.

This leaflet gives an overview of why behavioural challenges occur in PWS, as well as suggestions on how to manage them. However, an in-depth knowledge of your son or daughter is required to know how and when to apply different types of management techniques.

It is important to remember that, while this leaflet focuses on behavioural challenges in PWS, people with the syndrome have many positive characteristics which often offset their difficulties to a large extent.

They are known to be friendly, sociable, kind and caring, and many have a wonderful sense of humour. Each individual will also have other traits which are equally positive. Good behavioural management can bring these positive traits to the fore even more.



Common types of behavioural difficulties in PWS

- **Temper outbursts (tantrums, rages)**
- **Food seeking and/or stealing**
- **Stubbornness and resistance to change, argumentativeness**
- **Perseveration (asking the same question, or reverting to the same subject over and over again)**
- **Compulsive behaviour and insistence on routine**
- **Attention-seeking and manipulative behaviour**
- **Skin-picking**
- **Lying and blame-shifting**
- **Behavioural difficulties associated with mental health problems**

Factors influencing behaviour in Prader-Willi Syndrome

As with every other form of behavioural difficulty, not just those in PWS, there are many factors which are involved. These may include cultural and social influences, as well as parental or carer attitudes. At the other end of the scale, other genetic factors, apart from the PWS, may have some effect on the individual. More rarely, changes in behaviour and emotions can be the result of a mental health problem such as depression. However, there are certain elements which arise directly from the effects of PWS, and should always be taken into account when managing behaviour.



Problems arising from imbalance in the hypothalamus

Our understanding of PWS leads us to believe that these effects almost all arise from some kind of dysfunction or chemical imbalance within the hypothalamus region in the brain.

Although some progress has been made in finding medications which will help with some aspects of these imbalances, we still have a lot to learn, and there is no "magic bullet" for any aspect of behaviour in PWS. It is important to bear in mind that certain aspects of PWS behaviour stem directly from physical disturbances in the brain, and may not necessarily occur because the individual with PWS is being "naughty", "bad" or "difficult".

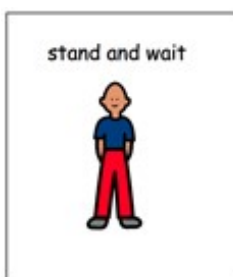
These physical disturbances may result in:

- Feelings of hunger, even after a meal, and thus the drive to eat and to seek out food
- Immature social skills
- Arrested or delayed emotional development
- Lethargy, somnolence and/or disturbed sleeping patterns
- High pain threshold (difficulty in feeling pain)
- Poor body thermostat (ability to keep the body at the right temperature).



Note:

For ease of reading, the text refers to "he/him" but all the information is equally applicable to girls.



Specific emotional and learning disabilities

Many individuals also have specific emotional and learning disabilities which can impact on behaviour. These may be evident even where the person is functioning at a relatively high intellectual level. These include:

- Difficulties in processing spoken information, with greater skill at understanding visual cues. In other words, show your son or daughter how to do something or what you want them to do by means of pictures, signs or actions, rather than telling them.
- Speech and language disorders which can be frustrating for both the person with PWS and the listener. These vary considerably from one person to another. You can find out more about these in the article on **Speech and Language in Prader-Willi syndrome**. <https://www.pwsa.co.uk/assets/files/speech-adults.pdf>
- Difficulties with short term memory, but a good long term memory; ie they may forget something learned only a few days or even minutes or hours before, but can remember significant events, such as journeys and celebrations, from a long time ago, and in quite a lot of detail.

Specific emotional and learning disabilities (continued)

- Mood swings, going from happiness to anger or sadness in a very short time period, sometimes triggered by relatively trivial events.
- Difficulties in appreciating that what happens in one situation may or may not happen in another. This means that once a fact is learned, it is difficult for the individual to "unlearn" it (this is particularly problematic if the "fact" proves to have been incorrect) or to appreciate that it may not always apply.
- A rigid method of thinking, often in terms of "black and white" which makes grey areas, "don't knows", and abstract concepts difficult for the person to understand.



- Difficulties with understanding time and numbers; money and budgeting.

The attitudes of other people

The attitudes of other people are also very important.

Likely to have a **negative** effect on a person with PWS's behaviour are:

- Aggressive attitudes and tones of voice
- A confrontational approach
- Being overly sympathetic
- Being in conflict with another parent/carer/relation - both giving different rules to the person with PWS or arguing in front of him
- Saying "Don't" as a means of trying to stop him doing something.

Likely to have a **positive** effect are:

- Firm, but loving and caring attitudes
- Ability to keep a sense of humour, and to see and exploit the funny side of a situation
- Ability to give clear guidelines and boundaries and stick to them
- Praise, and stressing the positive aspects of the person with PWS
- Saying "Do" as a way of showing him how to do something correctly, (ie instead of saying "Don't leave the door open", say "Please shut the door").



Behaviour management

What can make behaviour worse

A number of factors can make these behaviours worse, such as:

- **Major changes in daily routine or environment**
- **Changes in diet or meal routine**
- **Conflicts of opinion amongst others in close relationship to the person with PWS**
- **Unexpected happenings or upsets**
- **Major life stresses such as bullying, teasing, bereavement, moving residence, changing schools etc.**

On a daily basis, very minor occurrences can affect behaviour. For example:

- **Proximity to meal times**
- **Tiredness**
- **Minor change to routine**
- **Change in room or air temperature**
- **Mild teasing**
- **The person being unable to get their own way**
- **"Trigger" words or situations which are individual to your son or daughter.**

Managing behaviour



Bearing all the factors in the previous sections in mind, the following sections are offered as suggestions to help manage behaviour. Not all will work with everyone, nor all the time. Usually it is necessary to be always "one step ahead".

Some behaviours will be reinforced if they result in your son or daughter getting what they want. Thus your son or daughter may "act up" in one environment where his or her behaviour is "rewarded" by others "giving in", but not exhibit that behaviour in another environment where others take a firmer stance.

Quite often behaviours are very different between home and other environments such as day centres, colleges and residential care, which may reflect how your son or daughter is reacting to the people around them and the environment they are in.

Many people have noticed that individuals with PWS experience genuine remorse after an outburst of behaviour, and are really sorry for what happened, but this does not seem to prevent them from doing it again.

Communication

Problems may arise simply because your son or daughter has not fully understood the information which has been given them. If they have additional problems with short term memory, parts of complex sentences may be forgotten. Any information being passed needs to be broken down into small sentences or phrases to enable them to absorb completely what is being said. Even if your son or daughter's intellectual ability is apparently relatively high, this may still be the case. Also, those with greater intellectual ability may be unable to use their knowledge effectively within educational, social or work situations, due to immature social and emotional skills.



Temper outbursts

Temper outbursts, sometimes described as "tantrums" or "rages" are probably linked to the immature emotional development of people with PWS, and share many similarities with the type of tempers displayed by most two to three year olds, or by those going through puberty. Just as some little children and teenagers seem more prone to tantrums than others, people with PWS also vary considerably in the type and degree of tempers. It is very difficult to say why this variation occurs, but is most likely a mixture of all the influences which were previously mentioned.

In any one individual, tempers may increase or decrease over time, depending on circumstances. The years between 12 and 25 may be more difficult (as with many youngsters without PWS). Temper outbursts can manifest in any of the following ways, from fairly mild to severe:



- Screaming
- Swearing
- Throwing things
- Hurting themselves
- Crying at length, more often occurring at the end of an outburst.
- Being aggressive and violent towards other people.

Things that may trigger a temper outburst

Researchers have found that people with PWS are more prone to temper outbursts than people without PWS at similar levels of development, and that they are more vulnerable to having a temper outburst, as are others without PWS who have hypothalamic or limbic damage. (Limbic relates to the part of the brain concerned with basic emotions and instinctive actions).



This does not mean that people with PWS will always have outbursts, but that in certain situations they are more vulnerable to having them. Sometimes it is difficult to find a reason for the temper outbursts, but the most common "triggers" are listed below.

- Being denied food
- Being unable to get their own way (not just over food)
- A change in routine
- Someone else using their possessions
- Attention-seeking
- Tiredness
- Confusion or misunderstandings
- Low mood
- Teasing

There are several techniques which can be tried to help your son or daughter manage their outbursts. Personal knowledge of how different things affect them is essential.

Managing the environment to avoid outbursts



Many potential outbursts can be avoided by providing the right environment to suit your son or daughter's needs. This might include:

- A structured day, with plenty of advance warning if changes are planned.
- Unpressurised tasks or expectations. For example, don't expect your son or daughter to be ready to go out in five minutes if it normally takes them 20 minutes - give them plenty of warning, possibly using a "countdown" system, with a reminder at 15 minutes, then at 10 minutes, and so on. This is particularly important for those with higher intellectual levels who may be placed under greater pressure, because more is expected of them, but which they are often unable to deliver in the time space allowed. This includes workplace and educational tasks.
- If your son or daughter finds it difficult to be around food (most, but not all, do), remove food from sight, do not allow others to eat in front of him or her and, if necessary, lock food away.

Continued ...

Behaviour management

After an outburst

Identifying the cause of the outburst may determine how it is handled afterwards. Essentially, there are two main causes.

In the first instance, he has had the outburst because he cannot get his own way, wishes to divert attention away from something else he has done, or is attention-seeking. Once he has calmed down, it may be possible to talk very simply about your expectations about behaviour.

The second cause of outburst occurs when something externally has upset him: an unexpected change to routine, teasing or bullying, or a disappointment. Again, support can be given to your child to help him overcome outbursts in the future, but this can be in a more sympathetic manner - just as you would console anyone else who has been upset by a circumstance beyond their control.



Managing the environment to avoid outbursts (continued)

- Ensuring that all who come into contact with your son or daughter are giving the same message about what they expect in the way of behaviour.
- Help your son or daughter to learn techniques to keep calm such as deep breathing, listening to music, using worry beads or stress balls.
- Put up notices and pictures to remind your son or daughter to "keep calm" and/or of what happens when during the day.
- It may be useful to discuss in advance with him or her situations they find difficult and how to cope with the situation.

Look for signs of imminent outbursts

Most parents and carers who are in daily contact with the individual with PWS will recognise signs of increasing agitation, which vary from one person to another - sometimes it can be something as small as a slight quivering of the mouth, or a single tear rolling down the cheek. This is the time to try to head off the outburst before the storm begins to gather force:



- Use a calm tone of voice. Try not to show by your tone that you are exasperated, angry or becoming impatient.
- Distract attention away from the subject or situation by doing or saying something unexpected (and preferably funny), or talking about another topic which you know is of interest to them.
- Praise your son or daughter for something they have done, or remind them how well they coped in a previous, similar, situation.
- Suggest they go to a quiet place for a few minutes to listen to relaxation tapes or soothing music (especially if you suspect tiredness is a factor).
- Don't "wind up" the situation, and don't put ideas into his head by saying things like, "Don't you dare start screaming now!" or "Stop acting like a baby!" It is often better to make a non-confrontational remark, or say nothing at all.
- Keep very calm and cool. If possible, bring someone else in to take over.
- Do not start an argument, and say as little as possible about the situation which has triggered the "wobble."



"Weathering the storm"

Sometimes, even though every effort has been made to prevent an outburst, one will still occur. Then it really is a case of "weathering the storm" and, once over, everything goes back to normal again.

Outbursts in children are usually easier to manage, but in teenagers and adults, there is the added complication that the general public tends to stare at the person.

The outburst can vary from prolonged screaming and shouting (anything from 10 minutes to 2 hours) to, in a very small minority, violent and aggressive behaviour. There are no quick and easy answers to managing an outburst, but the techniques on the next page may help to alleviate the situation.

Managing an outburst

- Remove your son or daughter from the situation if necessary or remove the audience. (This of course is not always possible.)
- Take any other action necessary to ensure his safety and that of others.
- Stay very calm, talk softly and as little as possible.
- If possible, get someone else to take over if you are the object of the outburst.
- Do not attempt to reason or argue with him, even if he is undoubtedly in the wrong.
- Do not give in to his demands.
- Try distraction and humour again.
- Help your son or daughter to regain control by inviting them to join in or help with a task or activity.



Behaviour management

Aggressive and violent behaviour

Specialist restraint techniques may be needed for extremely violent or aggressive people who will hurt themselves or others.

If this happens frequently and you have no training in self-protective 'breakaway' techniques or minimal restraint techniques, ask a professional for help (eg GP, social services, health service professional).

Further information
If your son or daughter is showing violent or aggressive tendencies, you can request an article about this from the PWSA Office or find it online at <https://www.pwsa.co.uk/assets/files/Aggressive-or-violent-behaviour.pdf>

Anger management techniques



Ways to help your son or daughter might include basic anger management techniques:

- Relaxation DVDs (although you may need to do the exercises with your son or daughter so they can understand what to do).
- Help your son or daughter to identify physical feelings associated with emotional states, so they can recognise in advance when he might lose control, and take an agreed course of action, such as going to their room.
- Ask him to identify "triggers" ie, what upsets him. You may find that the first answer he gives is not the real reason.
- Getting rid of angry feelings by hitting a cushion or similar object. Be aware that although this can help some teenagers, for others it may increase the anger and they will be unable to "switch it off".
- Design visual aids such as pictures and symbols to help remind your son or daughter what to do.
- Brisk walking or other physical activity
- Listening to music.
- Using hand-held computer games to take his mind off the situation.



Food seeking and/or stealing

This type of behaviour can be particularly difficult. It helps to understand it if one imagines how it feels to be very hungry indeed: irritable, irrational, desperate to eat something (sometimes anything), thinking about where to get food from, etc.

As far as we know, this is how people with PWS feel for a good deal of the time, as the mechanism which tells them they are full does not function very well.

In many, this seems to be an "addiction" to food, just as some people are addicted to alcohol or drugs, with similar outcomes if they do not get it.

However, this also varies from person to person, ranging from some who have only a relatively mild preoccupation with food, to others who will go to extraordinary lengths to obtain it.

Managing the environment

Managing the environment is an important aspect of addressing challenging food-seeking behaviour. Depending on the severity of the problem, the following techniques might be used:



- Keep the kitchen locked, or make it "off limits".
- Use locks and/or alarms on fridges and freezers.
- Keep dustbins and rubbish bins locked.
- Alert neighbours and shop-keepers to the problem, especially if stealing/begging food or money is a particular difficulty. The police may also need to be informed.
- Ensure others do not eat in front of your son or daughter.



Managing food stealing

Even with the most alert parents and carers, many people with PWS can still manage to access food, sometimes in the most devious of ways.

Excessive weight gain is an almost certain sign that extra food is being obtained in some way, and it is sometimes very difficult to spot the loophole in the system. Look for the less than obvious!

For instance, offers to run errands for tutors or carers, or requests to go to the toilet, may take your son or daughter conveniently past kitchens or delivery vans. College and day centre friends who have items in their lunch box they do not like may be happy to give them away.

If your son or daughter is caught taking forbidden food, the immediate outcome may be a temper outburst, and that may need to be dealt with first. However, there are other points and techniques which might be tried (see next page).

Continued ...

Managing food stealing (continued)



- If you say to the individual "Did you take that chocolate?" many will vehemently deny it, despite evidence such as wrappers in their pocket, or chocolate on mouth and fingers. It is better to say "I know that you took that chocolate" if a denial is likely. Make sure however that you are absolutely certain you are correct before you say things like this. You can then move on to dealing with the problem, rather than getting involved in a "No, I didn't", "Yes, you did" situation.
- Try to agree some form of contract with your son or daughter. This may take several forms, depending on circumstances. For instance, for someone who has only occasional lapses, the contract might be, "If your weight goes up, you will not be able to do cooking at home/college." For someone with more serious lapses, the contract might be a reward system for not stealing.
- Refer to the contract you have already made - always stick to what you have written down and agreed will be the consequence of food stealing.
- If your son or daughter has stolen food or money off someone else, insist it is paid back, and an apology is made.
- Opinions vary as to whether denying an item of food at the next meal (eg pudding) is viable or even humane. Much depends on the individual - some will agree to this, others will not. On the other hand, giving a special food treat or meal as a reward for not stealing can be an option, again depending on the individual.



Resistance to change, argumentativeness

Researchers have found that people with PWS find it very difficult to switch from one thing which is engaging their attention to another.

This leads them to appear stubborn and refusing to "move on". Added to this are the specific learning difficulties which make it difficult for the person to understand abstract concepts.

Usually, they are not being deliberately difficult - they are having genuine difficulties coping with a situation.

However, in some cases, attention-seeking or manipulativeness may also underlie this behaviour - knowledge of your own son or daughter is essential in this respect.

Continued ...

Behaviour management

Helping to understand

You may find it helpful to show your son or daughter the booklet "Our Way of Life" .

There is also a workbook available, aimed mainly at children and younger people, which may be helpful for those with limited understanding. "Let's Talk about PWS".

<https://www.pwsa.co.uk/about-pws/publications/>

Remember!

Bear in mind that food-stealing behaviour is the consequence of a physical urge, and is not pre-meditated "bad behaviour", and it is very difficult for your son or daughter to understand the concept of theft. Any efforts made by them to overcome this urge should be highly praised.

Resistance to change, argumentativeness — Managing the environment



Helping your son or daughter to make decisions and to expect change may contribute towards minimising stubborn behaviour and reducing resistance to change.

- If you know a request is likely to result in stubbornness, help your son or daughter over it by suggesting a reward if they comply. This can be a practical reward such as a trip to the cinema or magazine, or the promise of extra time spent with a favourite person or on a favourite activity.
- If you know a change is going to happen, try to present it in a positive light eg "a big surprise" or give control back to your son or daughter by saying, "such and such is going to happen ... can you help me by ..."
- Give known alternatives well in advance, "If the weather is wet, we will..." "If Mrs Jones is out, we will ..."
- Use visual aids – pictures and diagrams – to help them understand what you want them to do.

If a particular situation always results in stubbornness, try to find out why this is happening. Has something bad happened in this situation in the past? Does it put your son or daughter in contact with people or places they feel uncomfortable with? Sometimes the stubbornness may be a coping device on the part of the individual, who has genuine reasons why they do not want to do something, but cannot find a way to tell you. Sometimes they may not even be consciously aware of it themselves.

After an episode of resistance to change

It is sometimes possible to talk through what happened, and point out the problems which it incurred. Even if they were not dangerous, it may have made you late for an appointment or upset others. Try to find out why it happened, so that you can both discuss what to do next time.



Managing resistance to change

Sometimes your son or daughter may absolutely refuse to move or comply with your requests. In extreme situations, this may put them, and you, in personal danger. For instance, sitting down in the middle of the road and refusing to move. These are some ideas to deal with the situation, but creative thinking plays a large part in this one!

- Try to help your son or daughter onto the next step by suggesting a pleasurable activity to do next. However, care needs to be taken to avoid them taking advantage of this because they can begin to feel they are being rewarded every time they act up.
- Keep very calm. Don't yell or shout.
- Try to be lighthearted. Also be aware that they have lost control of the situation, and need to be helped to gain face again, eg, "If you don't come with me now and show me the way home, I might get lost, and then what would I do?"
- If safe to do so, ignore him or her, and go on with what you were doing, especially if you suspect they are attention seeking.

Perseveration (asking the same question, or reverting to the same subject over and over again)



This type of behaviour can often be noticed in very young children, who continually say "Why?" after every question. In PWS it may be linked to this stage of emotional development, as well as their specific learning difficulties (eg auditory processing difficulties) and language difficulties. It can become extremely wearing to the listener. Very often the subject returned to will be food or meal-times. Here are some suggested techniques to help with this:

- Ask your son or daughter to repeat back what you have said to them, and then reassure them that their facts are correct.
- If it is a certain topic which is being fixated on, tell your son or daughter that they have x minutes more to talk about it, and then you are going to change the subject. If necessary, show them when they will have to stop by pointing to a clock.
- Do not continue to argue or reason with him or her. This will only add to their confusion.
- Divert the topic of conversation onto something else.
- If the answer to their question is genuinely "I don't know", you may need to explain why you don't know. Otherwise they may think that if they keep asking, sooner or later you will know the answer. They may also think they are just being fobbed off.
- Try to draw your answer so that he or she can see it visually.
- Be aware that if you keep answering the question every time it is asked, this may constitute a "reward" to your son or daughter, in that they are getting your attention. They may continue just to get the response from you.
- If they still continue with the subject, say you have heard enough and walk away.



Managing obsessive behaviour

- Provide a greater range of activities and things to do, especially social activities and meeting new people.
- Try to channel it into more positive activities eg craftwork.
- With staff at colleges, day centres and residential homes, change the key worker regularly, or use several people as key support staff. Ensure your son or daughter knows you will be doing this in advance.

Continued ...

Behaviour management

Obsessive-compulsive behaviour

Researchers have found that this type of behaviour is more common in people with PWS than in the general population.

Some believe that it is due more to developmental delay (being behaviour which is observed in young children); others that it is related to autism spectrum disorder or mental health problems.

It can be on a relatively minor scale, such as collecting brochures or doing things in a particular order, but it can become more serious if the obsession is centred on another person - this may often be a member of staff at college, day centre or residential home, or some-one who the person thinks they are "in love" with.

Other types of obsession may be with objects and picking at clothing.

Skin picking, which is very common, is addressed on the next page.

Managing obsessive behaviour (continued)



- Reduce the opportunities to be with the person (or thing) they are obsessed with, and support the person who is the subject of the obsession to appreciate that they may need to remove themselves physically to assist in diversionary tactics.
- Suggest that hoarded objects are sold at a car boot sale to make money for a special treat.
- Try not to say things like "Don't spend so much time with ..."
- Have another person present who gives him or her a lot of attention to divert attention away from the person they are obsessed with.
- Use time spent with the favoured person as a "reward" for good behaviour for not bothering them for the rest of the day, but limit the time to around 15-30 minutes.
- If obsessions are very severe, and are interfering with your son or daughter's quality of life by preventing them from getting on with their life, some specific types of medication may help. This should be given only with guidance from a psychiatrist or similarly qualified person.

Skin picking and spot picking

Skin picking or spot picking is very common in PWS. Like other behaviours, it varies in severity. Skin picking is often provoked by small spots and grazes, which are picked continually, and thus never allowed to heal. However, sometimes wounds are made where there was no wound previously. Any area of the body can be a target. Most common are the limbs and the head or face, but other areas may suffer. Some people with PWS have been found to indulge in picking at their rectums and genitals, and this should always be considered first if there are any bowel or other intimate problems occurring. Variations of this skin picking behaviour include pulling out hair, finger nails and toe nails.

It is thought that skin picking is actually a pleasurable activity for the person with PWS, as their high pain threshold does not warn them that the activity is becoming dangerous. Skin picking releases endorphins in the brain which make the person feel good, as well as enforcing the addictive nature of the habit. It often occurs when the person is bored, watching television, or anxious. Although it is often more distressing to the onlooker than the person themselves, skin picking does present serious problems because of the risk of infection into the wound.

Managing skin picking



- Keep your son or daughter's hands busy with small toys or objects to handle, hand-held computer games, some form of craftwork, rolling wool into balls, or use worry beads, stress balls, or kush balls.
- Teach your son or daughter basic first aid, the need to keep sores bandaged and to apply antiseptic ointment. Note that in a few cases, care must be taken that the ointment is not eaten.
- Negotiate a reward for not picking the wound, and give the reward when it is fully healed.
- In conjunction with the reward system, use visual aids (eg ticks/stars for every time picking does not occur in each day) to show how picking is decreasing. Reward "along the way" eg, when three ticks/stars are gained, a token is given, and when an agreed number of tokens are collected, they can be "cashed in" for an agreed reward.

Managing skin-picking (continued)

- Massage or stroke your son or daughter's arms, hands, feet, legs, head etc for 15 minutes each day.
- Be aware that an increase in skin picking behaviour can be a sign of anxiety about some other area of life. Take time to allow your son or daughter to talk through any worries they may have. Anxiety-management techniques may help.
- If related to boredom, provide more activities, or negotiate a range of activities your son or daughter could engage in more independently.
- Keep finger nails short to avoid infection.



Behaviour management

Managing manipulative behaviour

- **Maintain firm boundaries about what behaviour is acceptable, with good communication between all parties so that everyone is clear what has been said to your son or daughter when negotiating the boundaries.**
- **Use a written care plan or contract so that everyone "knows the rules".**
- **Reward or praise good behaviour.**
- **When communication does break down, do not focus on blame or recriminations - this will foster an atmosphere in which manipulative behaviour will flourish. Try to work out how to respond more consistently in future.**

Attention-seeking and manipulative behaviour

Many people with PWS appear to be very self-centred, and have little understanding of how what they do affects others. It is very much the type of "me-first" attitude which is exhibited by small children. This can result in attention-seeking behaviour.

Another reason for this may be that the social environment is not very stimulating, and others are not showing much interest in the person with PWS. Or the only time others do respond is to tell the individual off if they have done something wrong - thus reinforcing the wrong behaviour.

On the other hand, people with PWS can have a very sophisticated knowledge of how to manipulate situations to their own advantage, and this should never be under-estimated. A common example is that of the individual who tells one person in authority that someone else in authority has allowed them to do something, eg have an extra biscuit, go out after lunch etc.

Some people with PWS will also try "emotional blackmail." For instance, they may tell someone who cares about them that they have nothing to do, in order to get their specific attention. It is always important to check with someone else to see if this is really the case.



Managing attention-seeking behaviour

- Promise 15-30 minutes of special time with a preferred person if your son or daughter does not manifest the attention seeking behaviour.
- Take time to include him or her in positive conversations.
- Negotiate a reward schedule for not engaging in the behaviour.
- Develop coping strategies with and for your son or daughter when attention is not available (eg an alternative activity or range of activities).
- Try to provide attention when behaviour is good, and make a special point of praising him or her then.



“Lying” and blame-shifting

Lying usually occurs when the person thinks they will get into trouble for having done something, or to “cover up” by denying that they did anything. There is also a tendency to blame someone else for what has happened. Remember that lying is quite an abstract concept, and many people with PWS may have real difficulties in recognising what they say as lying. If they say it to themselves, it *becomes* the truth to them.

Managing lying

- Do not agree with the lies, but it is unwise to be too confrontational, as this will just escalate your son or daughter’s sense of being out of control of the situation.
- Ensure good communication within the environment so that everyone knows as much as possible about what is happening on a daily basis, and will thus be able to spot more easily if your son or daughter is lying.
- Make sure your son or daughter knows when they will get into trouble for doing something, and when it may not be their fault. Sometimes he or she will lie or shift the blame just because they think they may be in trouble (eg losing a personal item).

The use of medication in severely challenging behaviour

Generally speaking, the use of medication is not the first option for severely challenging behaviour even when this may be linked to an underlying mental illness such as depression, for which changes in lifestyle and the introduction of new interests can be very effective. Only after every other avenue, technique or environmental change has been tried and failed should it be considered, and then in consultation with a psychiatrist or other consultant.



There has been some research in the USA into medications used in PWS, which has indicated that newer generation SSRIs (selective serotonin reuptake inhibitors) can be effective for some people. However, even then, there are other considerations which need to be taken into account:

- No single medication is universally effective in PWS. Whilst SSRIs worked very well with some people, others (as in the general population) have had negative reactions, both behaviourally and medically.
- Dosages need to be carefully monitored. Many people with PWS do not need the full dose of a medication, and respond better to a quarter to a half the normal dose. A cautious approach to dosages is used, starting off with a very low dose.

Other sources of help

Social services If you have not yet had any contact with your local social services department, and your son or daughter’s behaviour is becoming difficult to deal with, then it might be a good idea to contact them and ask for an assessment for both yourself (as a carer) and for your son or daughter. Local services vary, but there may well be services which can offer help with behaviour and/or provide respite.

The Challenging Behaviour Foundation (CBF) is an independent charity providing information, support and workshops around challenging behaviour associated with severe learning disabilities to families and professionals. All information and support is free to families. **Family support line: 0300 666 0126** www.challengingbehaviour.org.uk

Mental health problems

Research into mental health problems in PWS is still in its early stages, but there is evidence to suggest that mental health problems may be more common in PWS than in the general population. Significant increase or changes in challenging behaviour should be considered as possible mental health problems. These problems may take a variety of forms:

- Depression
- Lethargy (extremely withdrawn and refusing to take part in normal social activities or even to get out of bed)
- Extreme mood swings
- Severe anxiety
- Acute psychotic episodes (feelings of persecution resulting in bizarre behaviour, often with extreme anxiety, hallucinations and mood abnormalities). These episodes have been noted to be somewhat different from the usual forms of psychosis and often occur in a cyclical form. Researchers have found that there seems to be a greater tendency to psychosis in those who have the disomy form of PWS, although it can also occur in the deletion form. You can find out more about psychosis on our website at

<https://www.pwsa.co.uk/pdfs/mental-health-with-specific-focus-on-psychotic-illness>

Most mental health problems in PWS can be helped with medication, bearing in mind the need for careful dosage, and the advice of a psychiatrist should be sought. Other strategies may also be helpful. Exercise has a beneficial effect on mood. Some types of depression may be helped by psychological techniques such as cognitive behavioural therapy, where negative thoughts are challenged.

PWS and autistic spectrum disorder

“True” autistic spectrum disorder (ASD) is found in a small minority of people with PWS—see <https://www.pwsausa.org/wp-content/uploads/2017/06/Autism-With-PWS.pdf>

However, many people with PWS show some of the characteristics of ASD such as obsessive behaviour, dislike of loud noises and resistance to change. In these instances, children respond well to management techniques used in ASD.

Other behavioural difficulties

This article has focussed on the main behavioural difficulties which affect people with PWS, but there are some behaviours which are less commonly associated with PWS, which may be manifested, eg attention-deficit disorders and autistic spectrum disorders. Specialist advice should be obtained for these.

Behaviour management

YouTube resources about managing behaviour in PWS

Behaviour and mental health in PWS - Tony Holland (IPWSO and PWSA Ireland)
Behaviour and mental health in PWS - Tony Holland <https://www.youtube.com/watch?v=5MfuGXRYSKY>

Unique Behavioral Features of Prader-Willi Syndrome—Elizabeth Roof (USA) <https://www.youtube.com/watch?v=98GvGJ9DaCo&t=2s>

Prader-Willi, Understanding and Managing Behaviour Part 1—Janice Forster (USA) <https://www.youtube.com/watch?v=OfF4e6Elmh8>

Understanding and reducing problematic behaviours in persons with Prader Willi syndrome - Kate Woodcock <https://www.youtube.com/watch?v=9ldH1HiSDpo&t=330s>



Further information from the International PWS Organisation

If you would like a hard copy of any of the articles below, please contact the PWSA office

Story-telling in PWS

https://docs.wixstatic.com/ugd/a71d4c_beb14ea282e8415d8339ca739a37284e.pdf

Coping with change

https://docs.wixstatic.com/ugd/a71d4c_8a2d60002e7aa4637ae9d67b6e5f518b.pdf

Managing a meltdown

https://docs.wixstatic.com/ugd/a71d4c_d5c6c19c85c444bf9b12099e1e8d9cbe.pdf

Skinpicking

https://docs.wixstatic.com/ugd/a71d4c_5385ce87c6e9b0d04423c273739003df.pdf

Setting boundaries

https://docs.wixstatic.com/ugd/a71d4c_e512389e05b64f53a11e6f9f0d742ce7.pdf

Promoting positive behaviour

https://docs.wixstatic.com/ugd/a71d4c_2c66429cb92144e884f6ffbbb195521f.pdf

Rituals and obsessions

https://docs.wixstatic.com/ugd/a71d4c_c0951d2448914c249427a5a2ec677dbb.pdf



On the positive side

Managing PWS can sometimes be wearing for all concerned, and a sense of humour is a must. Most people with PWS also have a wonderful sense of humour, which can be tapped in to and used to help them overcome some of their difficulties.

Because lack of control is such an important feature in your son or daughter's life (ie over food and emotions), whenever possible try to "give back" control by entrusting him or her with important tasks, praise whenever they do something right, or even just when they are feeling a little low. Positively develop and build their self-esteem.

Negotiation about behaviour with your son or daughter with PWS can have very positive outcomes. Things that others may consider insignificant maybe very important to him or her and sometimes it is relatively easy to reach a compromise by rewarding in very small ways.

Start each day anew. Your son or daughter will have mostly forgotten what happened yesterday, and so must you.

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