

Behaviour management in PWS

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 document gives an overview of why behavioural challenges occur in PWS, as well as suggestions on how to manage them. The suggestions for management are not necessarily related to the age of the child or adult, as each individual has a different level of ability, understanding, and social skills. An in-depth knowledge of the individual is required to know how and when to apply different types of management techniques.

The most common types of behavioural difficulties associated with PWS are:

- 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.

It is important to remember that, while this document 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.

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.

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 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 child or adult 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
- Delayed puberty and immature sexual awareness
- 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).

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 the person how to do something or what you want them to do by means of pictures, signs or actions, rather than telling them. Occasionally, the person with PWS may have difficulties processing visual information rather than spoken information, which could be highlighted by undertaking psychometric assessment.
- 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.
- 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.

- 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 number, money and budgetting.
- 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 person to "unlearn" it (this is particularly problematic if the "fact" proves to have been incorrect) or to appreciate that it may not always apply.
- Mood swings, going from happiness to anger or sadness in a very short time period, sometimes triggered by relatively trivial events.

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

The attitudes of other people are also very important. Likely to have a **negative** effect on the person with PWS's behaviour are:

- Aggressive attitudes and tones of voice
- A confrontational approach
- Being overly sympathetic
- Displaying dislike of the person
- Not talking to the person
- Talking to the person too much
- Being in conflict with another parent/carer/relation - both giving different rules to the person with PWS
- Saying "Don't" as a means of trying to stop a person 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
- The maintenance of positive social contacts with the person
- Praise, and stressing the positive aspects of the person with PWS
- Saying "Do" as a way of showing the person how to do something correctly, (ie instead of saying "Don't leave the door open", say "Please shut the door").

Managing behaviour

Bearing all the above factors in mind, the following 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 are reinforced if they result in the individual getting what they want. Thus a person 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. 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 the person with PWS 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 the person to absorb completely what is being said. Even if the person'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 five year olds. Just as some little children 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 also increase or decrease over time, depending on external circumstances. The years between 12 and 25 can be particularly 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
- Being aggressive and violent towards other people
- Crying at length, more often occurring at the end of an 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 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. (Again these vary considerably between individuals.)

- 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
- Teasing

There are several techniques which can be tried to help the person to manage their outbursts. Knowledge of the person and how different things affect them is essential.

Managing the environment

Many potential outbursts can be avoided by providing the right environment to suit the person'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 the person 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 work-place and educational tasks.
- If the person 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 the person and, if necessary, lock food away.
- Ensuring that all who come into contact with the individual with PWS are giving the same message about what they expect in the way of behaviour.

- Put up notices and pictures to remind the person to "keep calm" (see below) and/or of what happens when during the day.
- Help the person to learn techniques to keep calm such as deep breathing, listening to music, using worry beads or stress balls.
- It may be useful to write a list of things that the parent or carer knows will provoke an outburst, and look at ways of avoiding or minimising them in advance.
- It may be useful to discuss in advance with the person 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 the person.
- Praise the individual for something they have done, or remind them how well they coped in a previous, similar, situation.
- Suggest the person goes 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 their 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."

Managing an outburst

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. There is the possibility that those whose outbursts have been well-managed as children are less likely to have outbursts when they get older (although there is no research to prove this). The outburst can vary from prolonged screaming and shouting (anything from 10 minutes to 2 hours) to, in a minority, violent and aggressive behaviour.

There are no quick and easy answers to managing an outburst, but the following techniques can help to alleviate the situation:

- Remove the individual from the situation if necessary or remove the audience. (This of course is not always possible.)
- Take any other action necessary to ensure their 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 the person, even if they are undoubtedly in the wrong.
- Do not give in to their demands.
- Try distraction and humour again.

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).

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, the person has had the outburst because they cannot get their own way, they wish to divert attention away from something else they have done wrong, or they are attention-seeking. Once they have calmed down, it may be possible to talk through expectations about behaviour. Sometimes a contract can be agreed whereby good behaviour (ie not having an outburst) is rewarded.

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

Ways to help the person might include basic anger management techniques:

- Relaxation tapes (although remember that the person may need to be shown how to carry out the instructions on the tapes).
- Help the person to identify physical feelings associated with emotional states, so the person can recognise in advance when they might lose control, and take an agreed course of action, such as going to their room.
- Ask the person to identify "triggers" ie, what upsets them.
- Getting rid of angry feelings by hitting a cushion or similar object. Be aware that although this can help some people, for others it may increase the anger and they will then be unable to "switch it off".
- Design visual aids such as pictures and symbols to help remind people what to do.

- Brisk walking or other physical activity
- Listening to music.
- Using hand-held computer games to take their 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.
- Help the person to understand why they need to keep to a lower calorie diet and not get too overweight (a special pack for people with PWS is available free from the PWSA (UK) which might help with this).
- Ensure others do not eat in front of the person with PWS.

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, someone with no money who has a 200 yard morning walk to school or work may take milk from doorsteps. Offers to run errands for teachers or carers, or requests to go to the toilet, may take the person conveniently past kitchens or delivery vans. School friends who have items in their lunch box they do not like may be happy to give them away. If the person is caught stealing or obtaining 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:

- Try to agree some form of contract with the person. 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/school." For someone with more serious lapses, the contract might be a reward system for not stealing.
- 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 or keeping weight within limits can be an option, again depending on the individual.
- If you say to the person "Did you take that chocolate?" many will vehemently deny it, despite evidence such as wrappers in their pocket, or chocolate on mouths 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.
- 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 the person has stolen food or money off someone else, insist it is paid back, and an apology is made.
- Bear in mind that this behaviour is the consequence of a physical urge, and is not pre-meditated "bad behaviour", and it is very difficult for the person to understand the concept of theft. Any efforts made by the person to overcome this urge should be highly praised.

Stubbornness and resistance to change, argumentativeness

Stubbornness, resistance to change, and a tendency to be very argumentative are again reminiscent of the behaviour of a very young child, and may be linked to the person's emotional immaturity. Added to this are the specific learning difficulties which make it difficult for the person to understand abstract concepts, as well as their rigid ways of thinking. Usually, they are not being deliberately difficult - they are having genuine difficulties coping with a situation. However, in some cases, attention-seeking or manipulateness may also underlie this behaviour - knowledge of the individual is essential.

Managing the environment

Helping the person to make decisions and to expect change may contribute towards minimising stubborn behaviour and reducing resistance to change.

- Get young children used to the idea of choice by presenting simple alternatives as often as possible, eg "Would you like to wear a red jumper or a blue jumper today?" This may work with older people too.

- If you know a request is likely to result in stubbornness, help the person over it by suggesting a reward if they comply. This can be a practical reward such as a toy 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 the person 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 ..." or "If the weather is wet, what would you like to do instead?"
- Use visual aids to help the person 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 the person in contact with people or places they feel uncomfortable with? Sometimes the stubbornness may be a coping device on the part of the person, 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.

Managing stubbornness

Sometimes a child or adult's stubbornness gets out of hand, and the person 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 the person onto the next step by suggesting a pleasurable activity to do next. However, care needs to be taken to avoid the person taking advantage of this because they 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 the person, and go on with what you were doing, especially if you suspect they are attention seeking.

After a stubborn episode

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.

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

This type of behaviour can often be noticed in 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 the person 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 the person 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 or watch.
- Do not continue to argue or reason with the person. 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 they can see it visually, and keep it with them to refer to.
- Try to find out how the person finds it easiest to understand information. For some this may be in small steps, for others it is better if information is presented as a whole.
- Be aware that if you keep answering the question every time it is asked, this may constitute a "reward" to the person, in that they are getting your attention. They may continue just to get the response from you.
- If the person still continues with the subject, say you have heard enough and walk away.

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 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, school or residential home, or someone who the person thinks they are "in love" with. Other types of obsession may be with writing and receiving letters, and picking at clothing. Skin picking, which is very common, is addressed in a separate section later.

Managing obsessive behaviour

- Provide a greater range of activities and things to do, especially social activities and meeting new people.
- Suggest that hoarded objects are sold at a car boot sale to make money for a special treat.
- Try to channel it into more positive activities eg weeding the garden, craftwork.

- With staff at schools, colleges, day centres and residential homes, change the keyworker regularly, or use several people as keyworkers. Ensure the person knows you will be doing this in advance.
- 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.
- Try not to say things like "Don't spend so much time with ..."
- Have another person present who gives the person with PWS 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 the person'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.

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 people are not showing much interest in the person. Or the only time others do respond is to tell the person 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 underestimated. 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 they do not manifest the attention seeking behaviour.
- Take time to include the person in positive conversations.
- Negotiate a reward schedule for not engaging in the behaviour.
- Develop coping strategies with and for the person 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 the person then.

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 the person 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.

Skin picking and spot picking

Skin picking or spot picking is very common in PWS. Sometimes it begins when the child is quite young, sometimes when they are much older. 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, the person with PWS may make wounds 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 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 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 person does not eat the ointment.
- 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.
- Parent or carer massages or strokes the person's skin 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 the person 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 the person could engage in independently.

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. Many small children exhibit very similar behaviour. 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 the person'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 when the person is lying.
- Monitor the person's activities to ensure that opportunities do not arise which would trigger lying.
- Negotiate a contract with a person so that they are clear what would happen if they do lie.
- Make sure the person knows when they will get into trouble for doing something, and when it may not be their fault. Sometimes the person 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, unless this a symptom of an underlying illness such as depression. 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. If it is used, it is often to facilitate other behavioural management techniques. It is only rarely used in young children.

There has been some research in the USA into medications used in PWS, which has indicated that newer generation SSRIs (selective serotonin reuptake inhibitors), such as Prozac, 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 Prozac has 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.

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. It is not clear whether these are caused by outside triggers, low self esteem, or an inherent disposition brought on by PWS, or all three.

Mental health 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)
- Hallucinations and hearing voices
- 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. Some researchers have found that there may be a greater tendency to psychosis in those who have the disomy form of PWS.

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.

Other behavioural difficulties

This leaflet 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.

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 their lives (ie over food and emotions), whenever possible try to "give back" control by entrusting them 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 the person with PWS can have very positive outcomes. Things that others may consider insignificant maybe very important to the person with PWS, and sometimes it is relatively easy to reach a compromise by rewarding in very small ways.

Simple things like foot massages, aromatherapy treatment and sharing an interest in clothes can all be of immense help.

Start each day anew. The person with PWS will have mostly forgotten what happened yesterday, and so must you.

Other organisations offering information about behaviour management

Challenging Behaviour Foundation Email network and family linking scheme for parents to share problems of challenging behaviour, regardless of disability. Also general advice and information.
www.thecbf.org.uk **Family Support Line:** 0845 602 7885

Mencap Challenging Behaviour service www.mencap.org.uk - **Family Support Worker** 0845 602 7885

Acknowledgments

With thanks to Dr Natasa Momcilovic, Dr David Clarke, Dr Harm Boer, Terri Dorman and Yvonne Wanstall for their comments and very helpful suggestions.

Thankyou

We are most grateful to the M&G Staff Charity Fund for their support for the revision and production of this document.

© PWSA UK