



“Coping With Change” in People with Prader-Willi syndrome

Most people with Prader-Willi syndrome (PWS) have difficulty coping with change. This varies from person to person and can be a real problem for those families affected. Change can be anything from a substitute teacher at school, to a different route being taken in a car trip or a new cup being used. Change can also occur suddenly and unexpectedly, as in an electrical blackout. A bad reaction to change may result in the person with PWS refusing to comply with requests, routines or plans and can quickly escalate into perseveration (repeated questions or comments), arguments and aggression. “Shutdown” is another typical response to the anxiety associated with change. These responses can be stressful for families as they often occur at the most inconvenient times.

Why do people with PWS have difficulty coping with change?

The problem is thought to be linked with the inability to “switch” attention from one thing to another. People with PWS generally find it more difficult to switch attention. Researchers from the UK have shown that the greater difficulty a person with PWS has in switching their attention from one thing to another, the more resistance they show to change. It is also known that people with PWS tend to prefer repetitive routines and often exhibit ritualistic or inflexible behaviours. Although varied in their reactions to change, they all feel “safe” with set schedules and expectations. If a change can be predicted in advance, it is possible to use strategies to avoid much of the anxiety that may otherwise result from the change occurring.

What works to avoid anxiety related to change

These are simple and practical strategies that help to minimise the reaction to *predicted* change:

- Discuss a back-up plan in advance. For example, you could agree to buy pears if there are no apples at the fruit shop. The fact that this is discussed in advance can prepare the person for a different situation to the one they are expecting.
- Thoroughly plan all details and double-check things with the person with PWS and with others. Occasionally the person with PWS has an expectation that you are not aware of. For example, they may expect that you will be stopping for a cup of coffee on the way home, however if you are not aware of that and drive directly home, that would be perceived by them as a change. So it is always helpful to go over the details of any plans, beforehand.
- Agree on some rules for an outing. For example, a rule that “If there is any yelling for any reason we will return home.” Before the outing remind the person of the rule, using positive terms eg “your best behaviour will mean we can finish the trip and won’t need to come home early”. A set consequence of bad behaviour can be hard to enforce at the time, however, going home once can stop a lot of problems in the future.
- Praise the person whenever change is accepted graciously.



- Use picture-boards or written routines. Any change to routine is added to the board in advance. For example, a schedule could be written for the week. Doctor's appointments or variations in the normal week can be written on it. If the person with PWS has any problems with the plans they can tell you in advance so, hopefully, any problems can be discussed and a solution can be negotiated.

What works to limit reaction to an unpredicted, last minute change of plans?

At times, unpredicted and unplanned change does occur. If you are fortunate enough to be able to respond quickly you may be able to reduce anxiety.

Below are some strategies that parents and carers have found successful in similar situations

- Give a solution before explaining the change. For example if a favourite coffee shop is closed for renovations, start by saying, "We are going to have coffee today at another coffee shop. That'll be different, won't it! We can ask them to make it just how you like. The coffee shop we like will be open again next week but it is closed today so we can't go there today." The last piece of information given in this example is the fact that the shop is closed and the fact that they will still get a nice coffee, comes first.
- Give a visual example of the problem or the change. For example if you were planning to move a cupboard into a room and it did not fit, showing a person with PWS the cupboard outside the door is better than trying to explain the problem in words. Simple sketches can at times be more easily understood than words.
- Be patient and calm as it may take some time for the change to be processed and then accepted.
- Try to point out the positive aspects that are involved in the change.
- Expressing shock at the change, yourself, and asking the person with PWS to help in the situation can diffuse their reaction to the change if they are given some responsibility within the event.

Managing change is an important part of managing the overall well-being of a person with PWS. Effective management of change can reduce anxiety and improve outcomes in other areas such as health and behaviour management.

Even 'good changes', such as a holiday, can be a source of stress for the person with PWS and need to be managed as a change to routine. Perseveration, increased anxiety and worry about what will happen while they are away and so on, are often seen. Not telling the person with PWS about something 'good' coming up for them is an option, but not always a good solution.

Working out changes with the person with PWS, so that a better result might be achieved next time, can be useful. For example: "Next time, if you think your cat has been in a fight and might have an abscess, do you think it would be better to go straight to the vet instead of trying to fix it yourself?" If the answer is 'yes', then

you can incorporate it into their guidelines – write it up on their pin-board, and make sure they know the new plan!

After an argument, or a ‘blow-out’, and when the person with PWS has become quiet and even remorseful, ask them what *you* can do to help them next time something like this upsets them. You may be surprised at their answers and often they can lead to positive results. For example: “Please leave me alone for a while,” “Please listen to me,” “Please don’t treat me like a child,” These sorts of agreements can often prevent a future argument or blow-out.

It’s all about compromise

As the person with PWS grows older, their behaviours may mellow. There is an expectation from the person themselves, as well as parents, siblings and family friends that they will be treated more and more like an adult. As parents and caregivers, we know this is not always possible, but we can sometimes be a little more flexible and trusting in certain areas. It is difficult to let go, and there will be times that the trust between you will be broken, but if you compromise, safely, often you can reach an agreement. As PWS lack of judgement and poor emotional control are not visible when they are calm and happy always prediscuss consequences.

Practising increased, safe responsibility can facilitate positive behavioural change.

For example: Carrying a set amount of money for a specific, preplanned event, can encourage self confidence and trust. However, success with this will take much preparation, explanation and negotiation. Having a “back-up” agreement to cover times when such a compromise does not work successfully is essential, such as: “If you spend the money instead of using it for what we’ve discussed it will be best for you not to take the money next time. I will then continue to organise the payment so you are not tempted to spend it on food.”

People with PWS vary in personality and cognitive ability. Some people can cope with more responsibility than others. If monetary responsibility is not necessary it is best avoided so as not to set the person up for failure.

A parent says: The main thing about compromise is that if you are going to *take something away* from a person, you must always give something back. *For example:* “Unfortunately I cannot let you keep a dog in this house because we really don’t have room. But, you can keep a bird in a cage/goldfish in a bowl, and we could always arrange for you to help walk some of the poor dogs at the SPCA who never get a chance to go out”.

Appealing to their own sense of judgment (“we don’t really have room”) and getting them to see that fact; giving something back (goldfish, or bird, for example) and appealing to their sense of importance, of doing something for others (walking someone else’s dog), seems to work.

Preparing people with PWS for change gives them the opportunity to cognitively and emotionally process what is to happen and how the change will effect them. It may take time and effort to prepare people for change but the results are usually worthwhile. However, if the person with PWS is stressed for a reason unknown to you or they are aware of anxiety you may be experiencing, difficult behaviour can occur despite your use of all of the above suggested strategies. Don’t feel like they or you have failed. Keep applying positive proactive strategies. In time, improvements will be noticed.