

Temper outbursts

Many people with PWS experience temper outbursts. We have identified several common triggers to temper outbursts. These triggers include a change to routine or expectation, receiving conflicting information and losing something. The frequency and duration of temper outbursts varies from person to person.

Our research has shown similarities in the sequence of behaviours and emotions during temper outbursts. Often emotional behaviours, such as crying and emotional vocalisations occur early in an outburst sequence, and are followed by quickly rising anger and more overt behaviours such as aggression. Outbursts are frequently followed by expressions of remorse and the emotional behaviours that indicate distress can also occur towards the end of an outburst sequence.

We have carried out a number of studies that have focused specifically on the temper outbursts that individuals can show following changes to their routines or their expectations (plans). We found that such temper outbursts are linked to experiencing a difficulty with change, which is a broader characteristic of behaviour that is commonly shown by individuals with PWS. Many people with PWS find it very difficult to deal with changes to routines or plans and – even if they do not show temper outbursts following such changes – often get upset following such changes.

We assessed the different types of brain processes in people with PWS that we expected – based on research in other groups of individuals – might be linked to experiencing a difficulty with change. We found that people with PWS seem to have a specific deficit in the brain process called “task switching” or “attention switching”.

Task switching is the brain process that allows us to switch from thinking about a set of stimuli or events in one way, to thinking about the same set of stimuli or events in a different way. So, for example, if we have a boiled egg in a shell and we have a teaspoon, we might start by using the teaspoon to crack open the egg and then move on to using it to eat the egg with. In this scenario, we would have to switch between thinking about the teaspoon as an implement heavy enough to crack the egg, and thinking about the spoon as a piece of cutlery that is appropriate to eat the egg with.

Not only do people with PWS find switching particularly difficult, but when they engage in switching, the brain functions in a different way compared to people who don't have PWS. We found that people with PWS who have greater difficulties with switching, also show more difficulty with change. In addition, we found that using a computer task to place demands on switching abilities with participants with PWS, triggers these individuals to show behaviours – such as arguing, questioning and ignoring requests – that form part of their characteristic sequence of behaviours that occur during a temper outbursts.

Our research therefore showed that two important characteristics that can come together to cause temper outbursts in people with PWS are a cognitive deficit in switching and a difficulty with change.

Following these studies, we have therefore tried to use this information to help us to develop intervention approaches that may help families to reduce the difficulties with temper outbursts shown by the people they care for with PWS.

We noticed that for some individuals with PWS, they showed very little or no upset following changes, unless those changes happened in routines that they had been accustomed to for a very long time. We know that routines are important for children growing up with PWS, particularly around food, to reduce anxiety that might be experienced about not knowing what is going to happen when. But if allowing a routine to be established for longer would result in people showing greater difficulty when we subsequently change that routine, then this would suggest that we should try to develop ways to help people with PWS grow up with the routines that they need with flexibility built in.

Our research used games that were completely novel to participants with PWS. Participants played these games for different lengths of time so that there were different durations of opportunity for the game's routines to become established. After the routines had been established, a series of changes were made to those routines. We found that as participants were exposed to the routines for longer, they showed more temper outburst behaviours and increased emotional arousal following changes to those routines. This research tells us that it is important for us to consider that including flexibility in the routines that people with PWS are exposed to may be an important way that we can help to reduce future difficulties with change. Future research in this area will be very important to inform us about how to do this in the best way.

Because we know that changes to routines can trigger temper outbursts, we also explored whether increasing the predictability of a change to routine would reduce the number of temper outbursts that follow changes. We developed an intervention strategy that used a picture card and verbal phrase to signal changes to routines whenever caregivers had prior notice that these changes would occur. Parents/carers/teachers used the strategy and recorded the number of temper outbursts that participants with PWS showed using a diary. We also set up some situations in which we could observe participants' behaviour following changes to routines that were not signalled by caregivers, compared to following changes that were signalled. Our observations suggested that for the particular changes we observed, using the signal made it easier for people with PWS to deal with the change, and led to less temper outburst behaviours. The diaries kept by caregivers showed that for the majority of people who used the strategy, less temper outbursts were shown compared to before the strategy was used. We would like to extend this research to investigate what happens when caregivers use this strategy over a longer period of time and with less one to one input from researchers.