

ATTENTION AND BEHAVIOUR IN CHILDREN WITH PRADER-WILLI SYNDROME KATE WOODCOCK, CHRIS OLIVER, GLYN HUMPHREYS

Why did we decide to look at attention and behaviour in children with PWS?

When we attended meetings of parents and carers of people with PWS, many people were concerned with the difficulties that they face because of challenging behaviour. Researchers, such as Tony Holland and Joyce Whittington at the University of Cambridge, have found that people with PWS may have temper outbursts, ask repetitive questions and hoard things, as well as insisting on things being the same in their everyday life [see the PWSA-UK website].

We wanted to try to understand why these challenging behaviours are sometimes seen in people with PWS. The first way we wanted to try to do this was by talking to parents and carers about when the behaviour happens. This would help us to make predictions about the reasons why the behaviour might occur. The second way we wanted to try to understand the behaviours was by thinking about the way people with PWS may think. Perhaps the genes that cause PWS also cause the central nervous system of people with PWS to develop in a particular way that makes certain thought processes more difficult. This difficulty with particular types of thought processes might help us to understand why some of the challenging behaviours occur.

Who did we see?

We visited 46 children with PWS and their parents or carers. These children were between 6 and 18 years old and the average age was 14 years. We know that 30 of the children that we saw had the deletion subtype of PWS and 8 had the uniparental disomy (UPD) subtype. We also visited 28 typically developing children and their parents or carers. These children were between 5 and 11 years old, and the average age was 8 years and 5 months. Because of the learning disability that people with PWS have, the developmental age of the two groups of children was quite similar and this means that differences between the groups are likely to be caused by something specific to PWS and not by the learning disability that is part of PWS

What did we do?

Talking to Parents and Caregivers:

We asked parents and carers to tell us about any repetitive behaviour that their child showed. When we talked about each type of behaviour we made sure that parents told us when it usually happened, what usually happened before and what usually happened after. We also asked parents to complete two questionnaires that measure repetitive behaviour.

Assessments with the Children:

The first way we looked at the children's thought processes was by asking them to play a game on the laptop computer. This game helped us to look at three different questions. These were:

- 1) **Do children find it difficult to switch their attention?** Switching attention is changing from thinking about something in one way to thinking about it in another way. For example if you were playing with a child asking them to build a tower with coloured blocks, and then you asked them to make a pattern with the blocks instead. In this case the child would need to switch from thinking about the blocks as bricks and building a tower with the blocks, to thinking of the blocks as colours and about making a pattern with the colours.
- 2) **Do children find it more difficult to show a particular response when they also have to block another possible way of responding?** An example of this in everyday life could come from using a teaspoon; sometimes it might be used for stirring hot drinks and other times it might be used for eating a yogurt. If someone is using the teaspoon to stir a hot drink they must also block the action of putting the spoon to their mouth that they would be used to doing when using the spoon to eat yogurt.

- 3) **Do children find it easier to concentrate on where things are than on what things look like?** Studies have shown that there are two different paths in the brain for processing visual information. One path deals with attention to colour and form and the other path deals with attention to where things are.

The second way we looked at children's attention was by using a series of tabletop games to ask the questions:

- 1) How easily can children concentrate on particular pieces of information in their environment while ignoring others?
- 2) How easily can children do two things at the same time?
- 3) How easily can children stop themselves from responding in a routine way during an ongoing activity?
- 4) How easily can children stop themselves from doing something that is quite automatic that they have practised a lot so that they can do something that is less automatic that they have not practised?

What did we find out from talking to the parents and carers?

One of the main difficulties that parents and carers told us about was that their child often had temper outbursts, and that that these usually followed a change in routine, or a change in an event that their child was expecting. Parents and carers also talked about the repetitive questions that their children asked. These questions were usually about the future and what is going to happen and children often asked more questions if something changed. This leads us to think that predictability is very important for children with PWS. Children with PWS can find difficult to cope with changes in their environment.

"...when he's getting up in the morning he has to get himself dressed in exactly the same way...you can't interfere with that or he'll start tantruming..."

"...he can get himself so frustrated and it just happens so many times at home...and it's really really difficult...it's like a mini explosion and his face goes bright red...he'll shout and slam doors..."

"...at the weekend coz he doesn't know what's coming up it's like he's not even enjoying things...he's just interested in "When's my snack?" "When's my break?" "When's it gonna finish?"

We compared parents and carers responses to the questionnaires on repetitive behaviour between the PWS group and the typically developing group of children. Children with PWS generally showed more behaviour that suggests that they like their environment to be predictable (like a preference for routine) and more repetitive questions.

What did we find out from doing assessments with the children?

We compared children with PWS to the typically developing children and found:

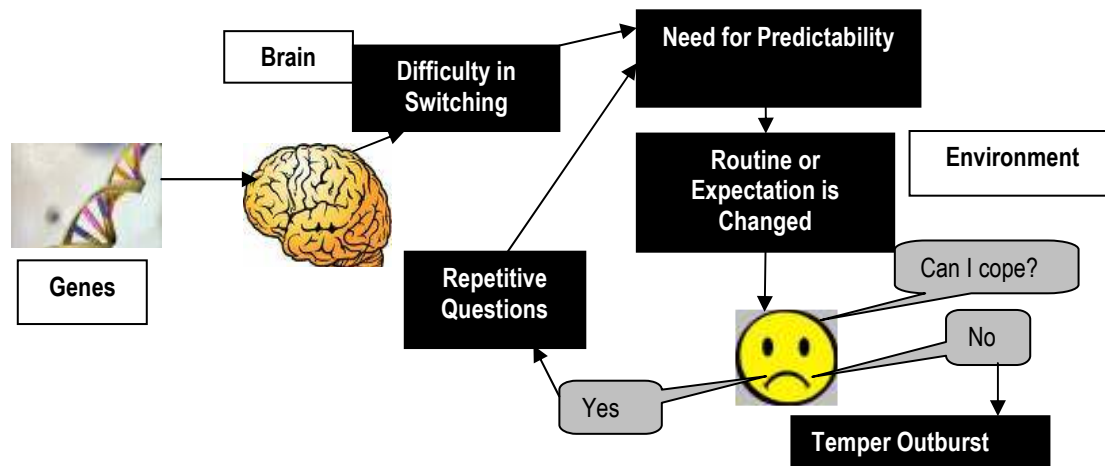
- 1) Children with PWS find it much more difficult to switch their attention.
- 2) Children with PWS find it much more difficult to do two things at the same time.
- 3) Children with PWS find it a bit more difficult to block one way of reacting when there are two possible things they could do.
- 4) Although children with PWS take longer to select particular pieces of information in the environment while ignoring others, they are very persistent and so very good at finding all of the information they are looking for.
- 5) Children with PWS find it easier to concentrate on what things look like than on where things are. This is the opposite pattern to typically developing children who find it much easier to concentrate on where things are.

How can we link what we found out from parents and carers with what we found out from doing the assessments with the children?

We wanted to see if the difficulty that the children with PWS showed in switching their attention was related to their need for predictability. We found that the children that showed a higher need for predictability in their environment (strongly preferring to have routines, to have

things done in a particular order or certain way and to have things in a certain place) also showed a greater difficulty in their ability to switch their attention.

The diagram below shows how the genes that cause PWS could interact with the development of people's central nervous system to cause a tendency to react to the environment in a particular way.



How could what we found out help in the future?

If we know that the temper outbursts shown by children with PWS are caused by their need for predictability then strategies that would increase the predictability in the children's environment may reduce temper outbursts. This could include things like timetables that show things that are going to happen. Many parents already employ strategies such as making sure that they try to give their children warning about something that is going to change.

If we know that it is a difficulty with switching attention that causes children with PWS to need their environment to be predictable then several strategies are possible that may reduce the children's need for predictability, and so reduce the temper outbursts that they show:

- 1) A method could be developed to teach children with PWS how to switch their attention so that they become better at doing this.
- 2) Children with PWS may be able to switch their attention more easily in certain types of environment. If this were the case we would be able to design ways to adapt children's environments so that they find it easier to switch their attention.

What are we going to do next?

We would like to be able to see if there are any differences in the behaviour and attention pattern that is shown in children with a deletion compared to children with UPD. This would help us to be more accurate when designing any education or intervention strategies. At the moment we have not been able to visit enough children with UPD to be able to do this comparison, so we would really like to visit some more children with UPD.

We also want to try to be sure that the deficit that children with PWS have in switching attention does cause them to need their environment to be predictable and that this need for predictability does cause temper outbursts when things change. We would like to do this by looking at some children's behaviour in much more detail. We would then be able to spend time with each child's family going through potential strategies that they could use to try to improve their child's behaviour. If you would like more information about this, please contact **Kate Woodcock** on **0121 414 3861**; parentfeedback@katewoodcock.com