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CEREBRA CENTRE FOR NEURODEVELOPMENTAL DISORDERS

TEMPER OUTBURSTS, REPETITIVE QUESTIONS AND ATTENTION IN CHILDREN WITH PRADER-WILLI SYNDROME

REPORT OF ASSESSMENTS CONDUCTED FOR RESEARCH

2007

We are happy to answer any questions and discuss any concerns you may have. Please do not hesitate to contact us.

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Results of Assessments conducted for Research Purposes
(This does not form a clinical assessment)

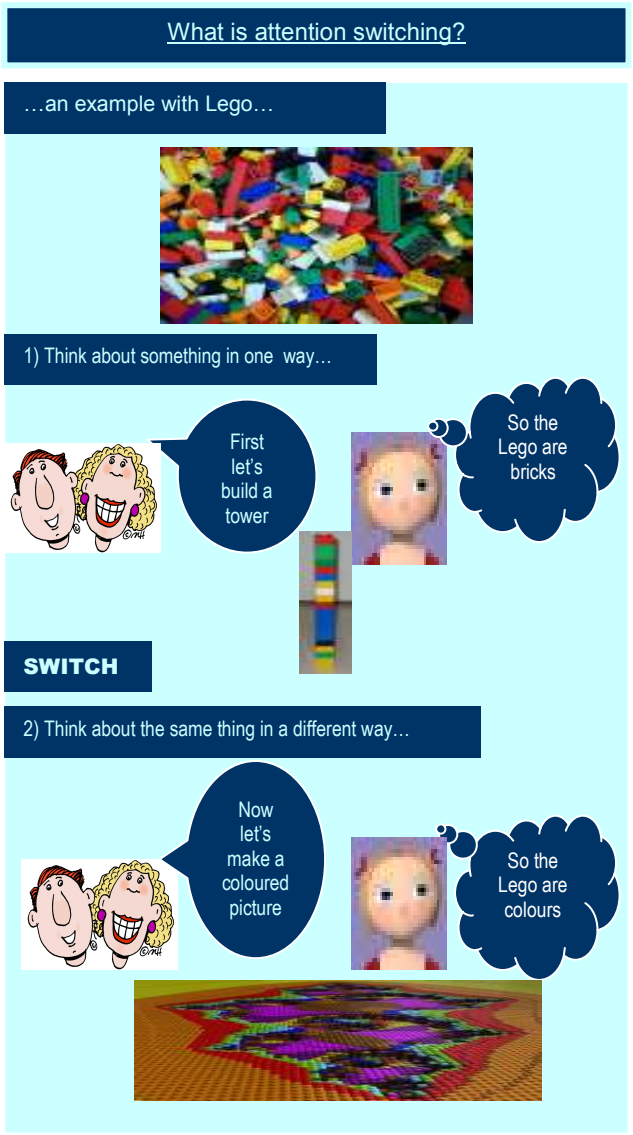
Project	Temper Outbursts, Repetitive Questions and Attention in Children with Prader-Willi Syndrome
Aims of Study	1) To investigate if problems with attention switching which we have shown to be present in children with PWS can give rise to repetitive questions and temper outbursts. 2) To look at the interaction between biological, cognitive, environmental and behavioural factors that may be involved in this process.
Dates of project	April 2007 – August 2007
Research Staff	Kate Woodcock, PhD student.
Name of Participant	(Anonymous) The name of the participant has been changed throughout.
DOB	
Address	
Diagnosis	
Parents/ guardians	
School	

Background Information

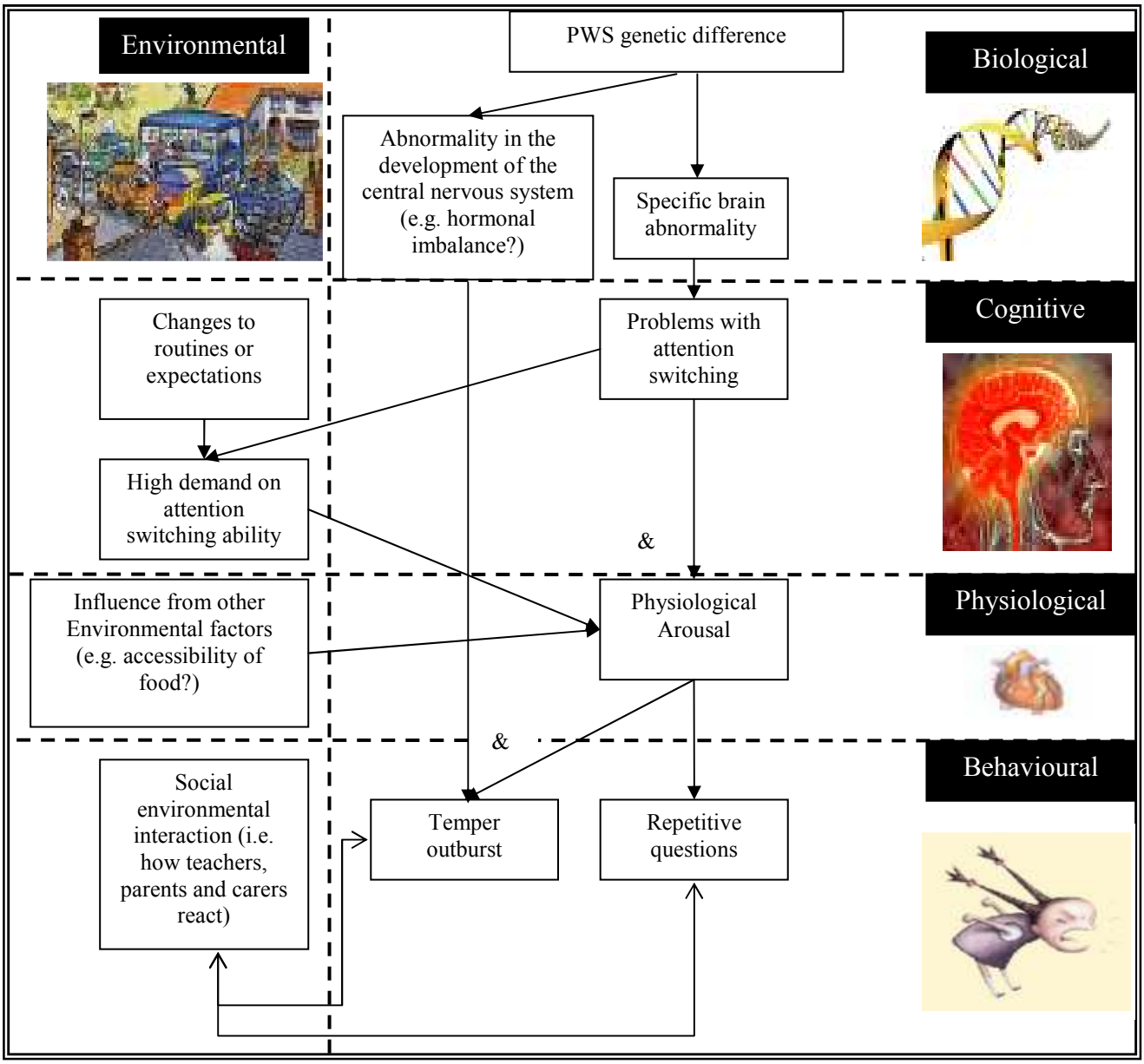
Results from the study on attention and behaviour in children with PWS, boys with Fragile-X syndrome and typically developing children that you and Jazmine participated in last year showed us that children with PWS seem to show particular difficulty in attention switching.

When we interviewed you and other parents and carers of children with PWS, you told us that changes to routines or expectations are often followed by children getting upset, asking more questions and / or having a temper outburst. We also found that children that showed more difficulty with attention switching also showed more preference for routine and repetitive questions.

This led us to think that maybe children with PWS prefer the same routine because changes to routines or expectations would mean that they have to switch their attention, which they find difficult. We drew a model that predicted that changes in routines or expectations place a high demand on children with PWS's ability to switch their attention, which in turn causes the children's physiological system to kick into action, increasing their arousal and making them feel uncomfortable. In order to make things more predictable and so escape from this uncomfortable state, children may ask questions, but sometimes the level of physiological arousal is just too high and so a temper outburst may occur.



The model drawn below shows how biological, cognitive, physiological and environmental factors might cause repetitive questions and temper outbursts in children with PWS. "&" signs show where more than one factor must interact to produce that effect



Aims of This Study
We wanted to investigate the relationship between the problem with attention switching shown in children with PWS, changes to routines and expectations, physiological arousal and repetitive questions and temper outbursts. We wanted to see if our results would provide support for the model we have described above. If this turned out to be the case then we would have a better idea about how to design coping and training strategies to reduce the difficult behaviour shown by children with PWS.

Stage 1: What we did
In this stage we wanted to see what would happen when we used specific cognitive tasks to change the level of the demand placed on Jazmine's ability to switch her attention. We designed four types of special computer game:

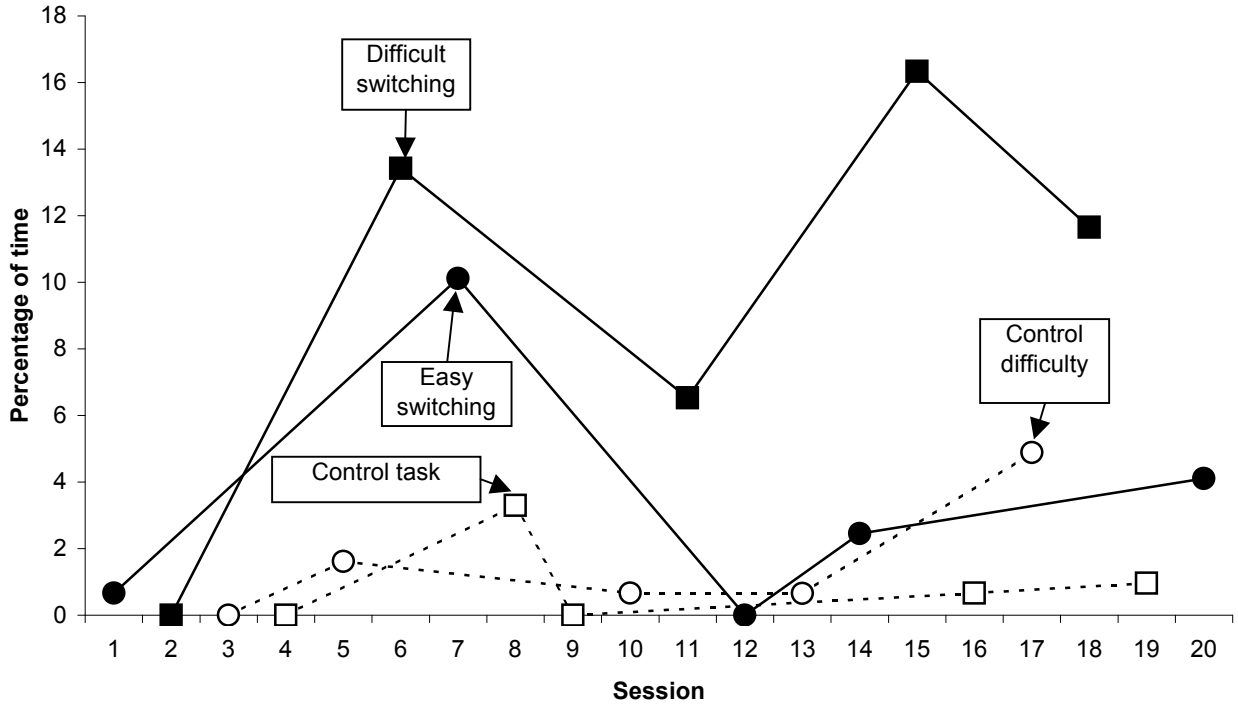
i. The difficult switching game placed a very high demand on Jazmine's attention switching ability.
ii. The easy switching game asked Jazmine to switch her attention, but a number of things were altered compared to the difficult switching game that we thought would make it easier for Jazmine to switch her attention. These alterations were: allowing more time for the switch, making the tasks that Jazmine must switch between more distinctive from each other and making it more predictable for Jazmine when she was going to have to switch her attention.
iii. The control task game didn't place any demand on attention switching, but it involved the same tasks as the switching games.
iv. The control difficulty game didn't place any demand on attention switching, but it was more difficult in terms of producing worse performance compared to all of the other games.

We asked Jazmine to compete five-minute long sessions of each of these games over and over again while we observed her behaviour using a video camera and measured her heart rate using a sensor attached to an elastic strap around her rib cage.

Stage 1: What we predicted
We predicted that Jazmine would show more repetitive questions and more of the kinds of behaviours that she shows during temper outbursts in the switching games compared to in the control games. This would tell us that these behaviours could be triggered by a demand on attention switching and were related specifically to a switching demand rather than a more general cognitive demand. We also predicted that the alterations that we made to reduce the demand on attention switching would reduce the questions and other behaviours shown during the easy switching game. This would suggest some ways that we might be able to make it easier for Jazmine to switch her attention. Finally, we predicted that measurements of Jazmine's heart rate would indicate that the demand on attention switching caused a state of physiological arousal.

Stage 1: What we found

This graph shows the percentage of time during each five-minute computer game session that Jazmine spent asking questions, frowning or arguing



Looking at the graph, we can see that across almost every session Jazmine showed more questioning, arguing and frowning during the difficult switching and easy switching games compared to the control games. This suggests that a demand on attention switching can cause Jazmine to show repetitive questions and behaviours that might occur during a temper outburst.

We can also see that Jazmine showed more questions, arguing and frowning in difficult switching games compared to easy switching games. This tells us that by allowing more time for the switch, making the tasks that Jazmine must switch between more distinctive from each other and making the switch more predictable for Jazmine, we could make it easier for her to switch her attention.

The measurements that we took of Jazmine's heart rate showed us that during the switching games she was more physiologically aroused compared to during the control games.

Stage 2: What we did

In this stage we wanted to see what would happen in a special environment that we were able to control, when we made changes to Jazmine's routines or expectations. We designed two types of special environment that we set up using games or activities that Jazmine was already used to doing or using new games that Jazmine had not played before:

i. The no-change environment allowed Jazmine to follow the routines that she was used to (e.g. in the pop-up pirates game each player used swords of only two colours) and everything occurred as she was expecting (e.g. if the expectation was set to stop playing when the alarm sounds, then that is what happened).
ii. In the change environment, changes were made to Jazmine's normal routines (e.g. in the pop-up pirates game players put in swords of any colour) or to her expectations (e.g. play did not stop when the alarm sounded even though Jazmine was given that expectation).

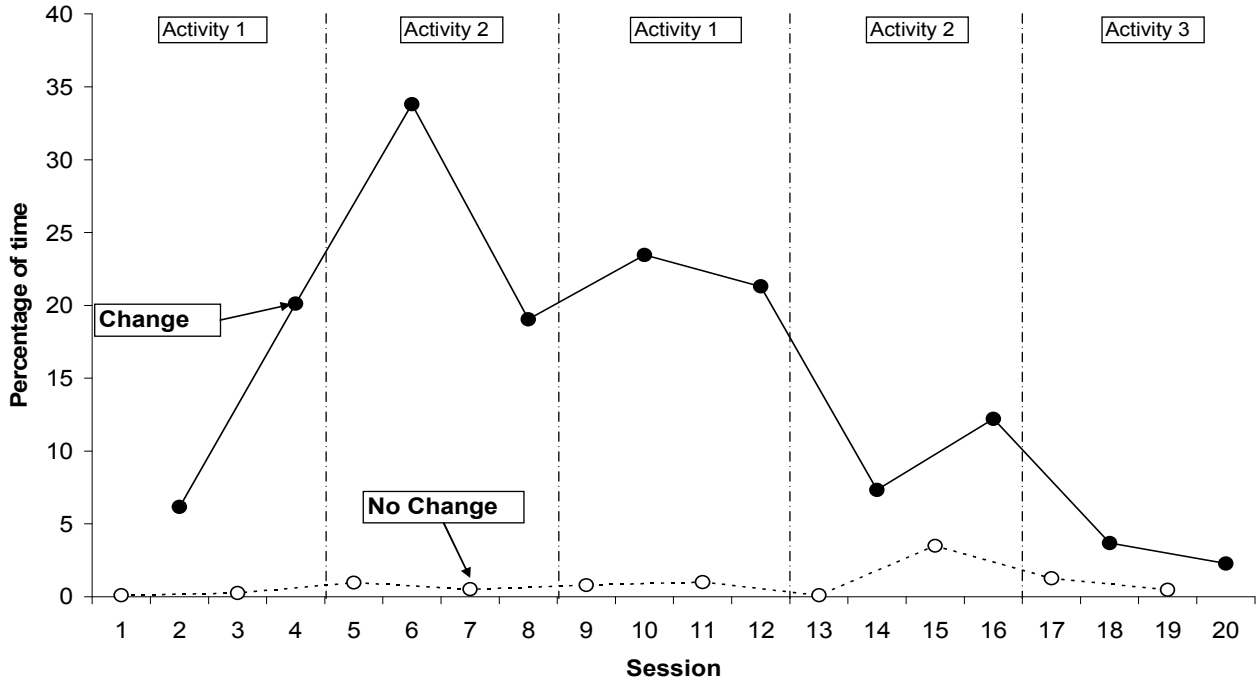
We asked Jazmine to compete five-minute long sessions in each of these environments over and over again using different activities, while we observed her behaviour and measured her heart rate.

Stage 2: What we predicted

We predicted that Jazmine would show more repetitive questions and more of the kinds of behaviours that she shows during temper outbursts in the change environment compared to in the no change environment. This would tell us that these behaviours could be triggered by a change in the environment (as well as by a demand on attention switching which we showed in stage 1). We also predicted that measurements of Jazmine's heart rate would indicate that the changes to the environment caused a state of physiological arousal.

Stage 2: What we found

<u>This graph shows the percentage of time during each five-minute session in each environment that Jazmine spent asking questions, frowning or arguing</u>



Looking at the graph, we can see that across every session Jazmine showed more questioning, arguing and frowning during the change environment sessions compared to the no-change environment sessions. This suggests that changes in the environment can cause Jazmine to show repetitive questions and behaviours that might occur during a temper outburst, as can a demand on attention switching.

Measurements of Jazmine's heart rate did not indicate that she showed higher arousal during change environment sessions compared to no-change environment, but other factors such as Jazmine's movements or breathing may have made it difficult for us to pick up such differences.

Stage 3: What we did

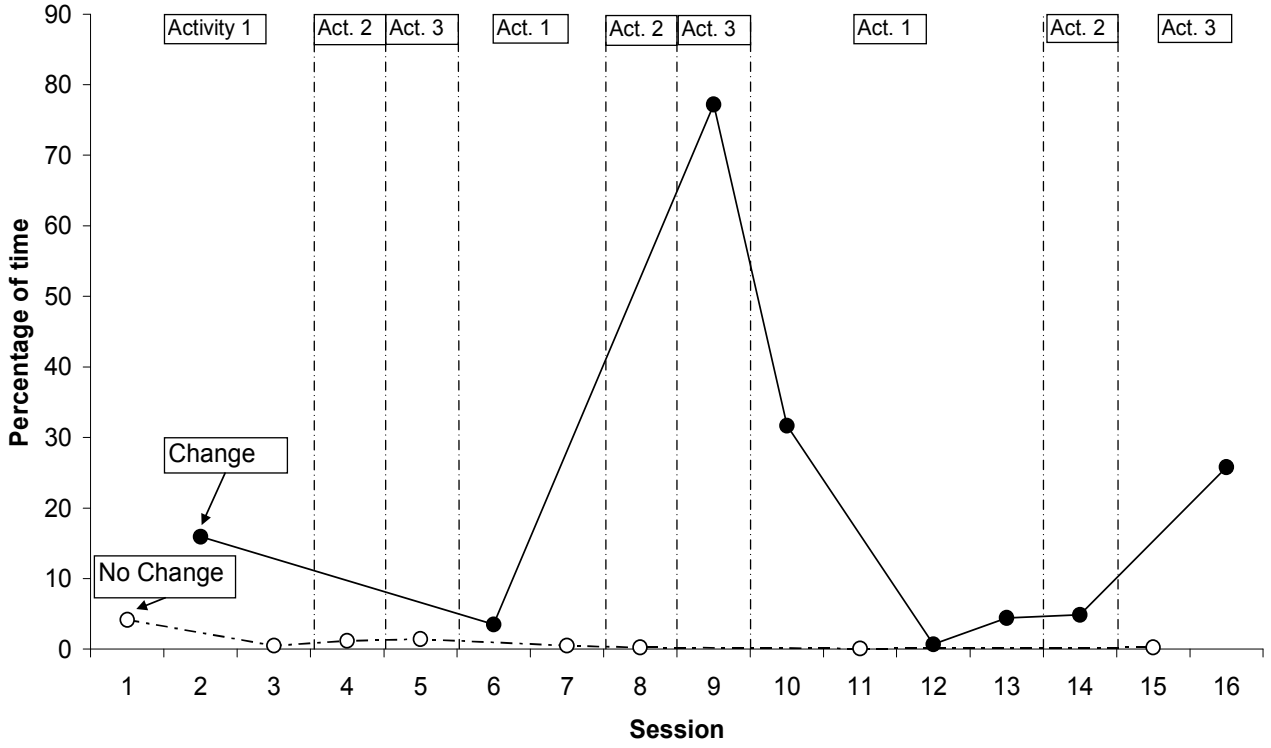
In this stage we wanted to see if changes in Jazmine's natural environment would cause her to ask repetitive questions and show temper outbursts. We observed Jazmine in her normal environment while she was doing a number of different activities. We observed each activity at least once when Jazmine was following her normal routine, or things happened as she had been told to expect (no-change). Then, at least once, we observed the same activity but where either Jazmine was asked to do something different to her normal routine or things did not occur as Jazmine has been told to expect (change).

Stage 3: What we predicted

We predicted that Jazmine would show more repetitive questions and more of the kinds of behaviours that she shows during temper outbursts, when routines or expectations were changed in her natural environment compared to when no changes occurred.

Stage 3: What we found

<u>This graph shows the percentage of time in each observation session in Jazmine's natural environment that she spent asking questions, frowning, arguing or crying.</u>



Looking at the graph, we can see that almost all of Jazmine's questioning, arguing, frowning and crying that occurred in the natural environment sessions that we observed, occurred when changes were made to Jazmine's routines or expectations. This tells us that changes to routines in Jazmine's natural environment can trigger her to ask repetitive questions and show temper outbursts.

What we found overall

So to summarise, we found that simply by placing a demand on Jazmine's attention switching ability using just a computer task, we could cause Jazmine's physiological arousal system to kick into action (this is what the heart rate measurements told us). Also, just from this computer task we could cause Jazmine to ask repetitive questions and show some of the behaviours (frowning and arguing) that she shows during temper outbursts.

Then, moving from a demand on attention switching that was not realistic at all, to a more realistic way that a demand on switching could occur; we showed that changes to routines or expectations could also cause Jazmine to ask repetitive questions and show some of the behaviours (frowning and arguing) that she shows during temper outbursts.

Finally we showed that in Jazmine's natural environment changes to routines or expectations were often followed by Jazmine asking questions or showing temper outbursts.

So it seems that at least some of the temper outbursts that Jazmine shows are likely to be caused by the problem that she has with switching her attention. In stage 1 we found a number of things that might make it easier for Jazmine to switch her attention and so it might be possible to translate these into ways that might make changes easier for Jazmine to deal with:

i. If Jazmine were given more time to accept changes, perhaps by being given more warning that a change will occur.
ii. If changes were made more predictable for Jazmine. This seems a bit difficult to translate into every day life, but perhaps if for example, Jazmine was told that she was going to do something new every Tuesday and then if on Tuesdays an activity that was fairly normal for Jazmine was carried out, but in a different way to usual. Then gradually it might be possible to change Tuesdays for a different day and then maybe just a particular time. So this would be like trying to make a routine of not having a routine.
iii. If when Jazmine had to change her routine or expectation, the environment around her was made somehow different to how it would usually be when she followed her normal routine. So for example, maybe if Jazmine were told that when someone was wearing a particular hat, it meant that things would happen differently to usual.

All of these ideas are just possibilities. We do not know from our research if they are all important, or which are the most important. This is something that we would like to look into more in the future.

As we have shown that some of the temper outbursts that Jazmine shows are likely to be caused by the problem that she has with switching her attention, this suggests the possibility that by training Jazmine to switch her attention better using a cognitive training programme, this might have a positive effect on Jazmine's behaviour. This is another project that we are currently working on and we would like to get back in touch when we have fully developed a training programme that Jazmine could try.

In many of the situations when we expected Jazmine to show really difficult behaviour, such as when we asked Jazmine's mum to change the content of her packed lunch, this behaviour did not occur. In this example, Jazmine noticed that she had one large apple instead of three small fruits when she was unpacking her lunch, so she took some plum tomatoes out of her salad and placed them in a row next to her apple and counted out loud "one, two, three". It seems that in this case something allowed Jazmine to get over the change. It might be that Jazmine's coping strategy meant that she did not actually have to switch her attention, because she continued to think that she had the usual three pieces of fruit for lunch, and in fact when I pointed out to Jazmine that she only had one piece of fruit, she argued with me. This illustrates the fact that many of the behaviours that Jazmine shows may be very useful for her and help her to get over some of the difficulties that she faces.

Although we have been able to show that some of Jazmine's temper outbursts can be caused by her difficulty with attention switching, this does not mean that that is the cause of all of her outbursts. Also, although we have identified changes to routines and expectations as one way that real life might be able to place demands on attention switching, there may be other ways. Generally however, we feel that we have made great progress with this study by showing that some of Jazmine's difficult behaviours can be directly linked to a specific difficulty that she has with the way that her brain is able to process and deal with certain demands.