THE RELATIONSHIP BETWEEN SPECIFIC COGNITIVE IMPAIRMENT AND BEHAVIOUR IN PRADER-WILLI SYNDROME

ABSTRACT

Background

Individuals with Prader-Willi syndrome have been shown to demonstrate a particular cognitive deficit in attention switching and high levels of preference for routine and temper outbursts. This study assesses whether a specific pathway between a cognitive deficit and behaviour via environmental interaction can exist in individuals with Prader-Willi syndrome (PWS).

Methods

Four individuals with PWS participated in a series of three single-case experiments including laboratory based and natural environment designs. Cognitive (computer based) challenges placed varying demands on attention switching or controlled for the cognitive demands of the tasks whilst placing no demands on switching. Unexpected changes to routines or expectations were presented in controlled games, or imposed on participants' natural environments and compared to control conditions during which no unexpected changes occurred. Behaviour was observed and heart rate was measured.

Results

Participants showed significantly increased temper outburst related behaviours during cognitive challenges that placed demands on attention switching, relative to the control cognitive challenges. Participants showed significantly increased temper outburst related behaviours when unexpected changes occurred in an experimental or the natural environment compared to when no changes occurred.

Conclusions

Difficult behaviours that could be triggered reliably in an individual by a specific cognitive demand could also be triggered via manipulation of the environment. Results suggest that a directional relationship between a specific cognitive deficit and behaviour, via environmental interaction, can exist in individuals with PWS.

Keywords: Prader-Willi syndrome; executive functioning; repetitive behaviour; attention switching; temper outbursts; preference for routine

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INTRODUCTION

Prader-Willi syndrome (PWS) has a prevalence rate of 1:10,000 to 1: 25,000 and is caused by absence of paternal genetic information in the chromosome 15 q11-q13 region either through a paternal deletion (60-70% of individuals), maternal uniparental disomy (UPD: 25-30% of individuals), chromosomal translocations or mutations of the imprinting centre (Boer et al., 2002). The physical phenotype includes distinct facial characteristics and short stature (e.g. Cassidy, 1997). Most individuals with PWS show a mild to moderate intellectual disability and relative strengths in academic achievement and visual processing (Dykens, 2002; Whittington et al., 2004 b), and weaknesses in auditory processing, mathematical skills and short-term memory (Bertella et al., 2005; Stauder, Brinkman & Curfs, 2002; Walley & Donaldson, 2005). PWS has been consistently associated with a behavioural phenotype that includes excessive eating, specific repetitive and self-injurious behaviours, temper outbursts, mood disturbance, excessive daytime sleepiness/under activity, and ‘disobedience’ or ‘stubbornness’ (e.g. Einfeld, Smith, Durvasula, Florio & Tonge, 1999; Richdale, Cotton & Hibbit, 1999; Holland et al., 2003; Moss, Oliver, Arron, Burbidge, & Berg, 2009).

It has been suggested that deficits in executive functioning (cognitive processes that allow the control and regulation of behaviour; Alvarez & Emory, 2006) may be associated with some aspects of the PWS behavioural phenotype (e.g. Wigren & Hansen, 2005). Executive functioning in PWS has been investigated by Stauder et al. (2005), who used a go/no-go task and measured event related potentials: they found that people with PWS showed an impairment in early modality specific inhibition compared to typically developing age matched controls. Jauregi et al., (2007) administered standardized measures of executive functioning that assessed word association, semantic fluency, mental flexibility and set-shifting. The scores of the individuals with PWS were significantly lower compared to those of normative populations. Some studies have used questionnaire measures to investigate potential links between executive dysfunction and behaviour in PWS, and have shown that higher executive dysfunction scores were associated with higher levels of repetitive and aberrant behaviours (Wigren & Hansen, 2005; Wally & Donaldson, 2005). However, these studies are limited in their ability to demonstrate a specific executive deficit in PWS over and above the global intellectual disability, or an association between performance on direct measures of executive function and behaviour.

We recently compared children with PWS to children with Fragile X syndrome and typically developing children on a task adapted from the Simon spatial interference task, which taps particular executive capacities (withheld for blind review). When controlling for the level of intellectual disability, the children with PWS showed a specific deficit in attention switching, which was associated with higher levels of preference for routine and predictability. In a related study, interviews with the parents and carers of children with PWS showed that changes to routines or expectations prompted emotional change, more repetitive questioning, and an increased likelihood of temper outbursts (withheld for blind review). On the basis of these two studies, we hypothesise that the deficit in attention switching in PWS can lead to a high demand on cognitive resources when the environment becomes less predictable. This cognitive demand in turn, can lead to an aversive state (possibly associated with high arousal) that would result in a resistance to change as individuals would be motivated to escape or avoid this punishing state. The cognitive demand and resultant aversive state may evoke repetitive questions (to reduce the cognitive demand or reinstate predictability), or temper outbursts if the aversive state persists (see withheld for blind review).

With this hypothesis, we do not mean to suggest that all temper outbursts shown by individuals with PWS occur as a result of a cognitive deficit in attention switching, or that all individuals with PWS and a deficit in switching will necessarily show temper outbursts. Instead, we hypothesise that a specific pathway between cognitive function and behaviour *can* exist in individuals with PWS. Such a pathway would have important implications for intervention and for informing associations between genes, the brain and behaviour.

In this study we will test the hypothesis by systematically manipulating the level of demand placed on the attention switching capacity of children with PWS and the level of predictability in their environment, while observing the effects on behaviour. By doing this we will evaluate the causal relationship between the deficit in attention switching, the resistance to change and the temper outbursts seen in PWS. Using multiple individual case designs, we will not attempt to investigate the specificity (or generalisability) of the discussed pathway between cognition and behaviour to individuals with PWS. Instead, we aim to investigate the viability of the mechanisms hypothesised to play a role in the relationship between a specific cognitive process and behaviour.

METHODS

Participants

Participants were recruited via a database of families of children with PWS who had agreed to be contacted. All participants had taken part in previous studies by the authors (withheld for blind review) and had been reported to show temper outbursts. Ethical approval was given by the Ethical Review Board at (withheld for blind review).

RB is female, and was 14: 2 years old at the beginning of the study. AH is female, aged 14:3 years; AG is female, aged 12:6 years; and KT is female, aged 17:1 years. All participants showed a deletion of chromosome 15 within the q11-13 region[1]. Approximately one year before initiation of the study, subtests from the Wechsler Intelligence Scales for Children (WISC III; Wechsler, Golombok & Rust, 1992) were administered to assess general cognitive functioning; items from the Repetitive behaviour Questionnaire (Moss & Oliver, 2008) and the Childhood Routines Inventory (Evans et al., 1997) were administered to assess preference for routine and repetitive questioning; and an adapted version of the Simon spatial interference task (Simon, 1967) was administered to assess attention switching. Parents or carers were asked to provide a description of the behaviours that occurred during a temper outburst shown by their child. These assessments are shown in Table 1.

[Table 1 about here]

[1] The genetic status of participants was not ascertained for the present study but had been confirmed with previous genetic testing.

Measures

Single case experimental designs allow an individual to act as their own control by exposing the individual to different carefully controlled conditions. This methodology is well established for assessing cognitive capacities in relation to external stimuli in brain injured patients (e.g. Riddoch & Humpreys, 1983) and for assessing behaviour in relation to the environment in individuals with intellectual disabilities (e.g. Iwata, Dorsey, Slifer, Bauman & Richman, 1982; Carr & Durand, 1985). Using single case experimental designs it has been possible to demonstrate a diverse range of highly specific triggers for challenging behaviour in certain individuals with intellectual disabilities (e.g. Murphy, MacDonald, Hall & Oliver, 2000). In this study we observed participants in conditions designed to manipulate the demand on attention switching and level of predictability in the environment within both highly controlled and more naturalistic settings in order to enhance social validity. In all cases, as is necessary to maintain experimental control, all experimental conditions were contrasted with tightly balanced corresponding control conditions.

Behaviour Observation

Across experiments each participant was observed for between five and eight hours and filmed using a video camera. Operationally defined behaviours were subsequently coded (continuous recording using ObsWin 3.2; Martin, Oliver & Hall, 1998). Two observers independently coded at least 10% of the footage for each participant and

Kappa reliability indices were calculated based on 5s intervals[2]. Behaviours coded were repetitive questions and behaviours observed that matched carers' descriptions of behaviours that their child showed during temper outbursts (see Table 1); definitions were: questioning (questions pertinent to the current situation), arguing (statement of disagreement, giving orders, making demands, or refusing requests), crying (crying with tears or speech or non-speech vocalizations associated with crying), frowning (lowered position of the inside ends of the eyebrows and a furrowing of the brow), stereotypical movement or vocalization (inter-weaving of the fingers with hands at midline in front of the mouth, tapping of the fingers on the forearm or non-speech vocalization from the throat), ignoring requests (not carrying out a direct request made by the researcher or a teacher before the request was repeated), mouth stereotypy (movement of the lips whilst not engaged in vocalization) posture changes (movement of the whole body to result in a different sitting position, or the raising and lowering of the shoulders coupled with lowering of the trunk), and looking at the researcher (eyes looking in the direction of the researcher). All Kappa indices indicated at least satisfactory reliability (> .4) with a mean value of .70.

Lag sequential analyses were used to identify clusters of temper outburst related behaviours. Within behaviour clusters, the conditional probability of a particular behaviour (target) being shown, given that another specific behaviour (criterion) would occur after a given time period or had occurred a given time period before, was higher than the unconditional probability of that target behaviour occurring due to chance

[2] Inter-observer reliability was calculated based on this proportion of observation time in order to ensure that the reliability indices were not falsely raised by agreement on non-occurrence of behaviour during sessions during which very low rates of the observed behaviour were shown.

alone (Bakeman & Gottman, 1997; Whitaker, Walker & McNally, 2004). To identify *temper outburst related behaviours*, criterion behaviours were set as the two most disruptive behaviours shown by each participant (crying/ ignoring and arguing). Target behaviours that showed an association with a Yules Q value of greater than .4 (equivalent to an Odds Ratio of 2.33) within a two-minute period preceding or following a criterion, formed the same behaviour cluster (withheld for blind review). behaviours that are controlled by the same contingencies (occur under the same antecedent and consequent conditions) can be viewed as functionally equivalent (DeLeon, Fisher, Herman & Crosland, 2000; Emerson, p. 79, 2001). The behaviour clusters (temper outburst related behaviours) identified for each individual therefore included behaviours that reliably occurred either during or preceding a temper outburst (see withheld for blind review). behaviour clusters consisted of crying, arguing, questioning and frowning for RB; crying, arguing and questioning for AH; crying, arguing, questioning and stereotypical movement or vocalization for AG; and ignoring requests, questioning, mouth stereotypy, looking at the researcher and posture changes for KT[3].

EXPERIMENT 1

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[3] All behavioural results reported below consider all temper outburst related behaviours for each participant in order to present the clearest possible description of the results. However, the same pattern of results was apparent when only the most disruptive behaviours shown by each participant were considered (those with highest face validity as temper outburst behaviours: arguing, crying, questioning and ignoring requests).

Experiment 1 examined the effect of a specific cognitive antecedent (high demand on attention switching) on repetitive questions and temper outbursts. In typically developing participants attention switching is often investigated using task switching paradigms (Rogers & Monsell, 1995) when a number of factors influence the ease of task switching. These are: a) when the stimuli are univalent (afford a single relevant response) compared to bivalent (e.g. Diamond, Carlson & Beck, 2005; Poulsen, Luu, Davey & Tucker, 2005), b) longer preparation time for the switch (e.g. Poulsen et al., 2005; Kray 2006; Lien, Ruthruff, Remington & Johnston, 2005) and c) increased predictability, for example through an additional external cue (e.g. Kray, 2006; Rubin & Koch, 2006).

To evaluate the effect of switching we designed four conditions to be administered using a series of computer games. These placed high or low demands on attention switching or controlled for the general cognitive demands of the tasks without placing any demand on switching. Importantly, by contrasting switching conditions to control conditions it was possible to separate any changes in behaviour that related to specific demands on attention switching from any that related to demands on more general cognitive resources. We predicted that when exposed to these conditions, participants would show more temper outburst related behaviours in the switching conditions relative to the control conditions and more of these behaviours in the high switching demand conditions relative to the low switching demand conditions.

Methods

Measures

Switching challenges

Four conditions were presented via a laptop computer each within the context of three different games (different sets of stimuli and tasks) in order to reduce practise effects. In all conditions a series of trials was presented that began with the presentation of a pictorial cue that filled the screen and indicated the task to be performed, which was to classify the stimulus based on shape or colour. A central stimulus was then presented (whilst the background cue remained) that required one of two response key (Z or M) alternatives. A visual feedback screen presented a central schematic face (happy for a correct response, sad for incorrect) following a response.

Difficult Switching condition (DS).

This condition placed a high demand on attention switching. Bivalent stimuli were presented following a short (500ms) presentation of the background cue. Stimuli were either congruent (50%; required the same response regardless of the task type) or incongruent (required a different response for each task). Following a response, the feedback screen was displayed for 1000ms. The two tasks were presented randomly so that approximately 50% of trials required a switch. Switches were therefore unpredictable for the participant.

Easy Switching condition (ES).

In this condition the difficult switching protocol was adapted to reduce the demands on attention switching. The purpose of this condition was twofold: 1) to assess whether factors affecting the ease of task switching by typically developing individuals would also affect the ease of switching in individuals with PWS and 2) as an additional method of control because this condition involved switching but was designed to place a lower demand on general cognitive resources than all of the other conditions. The stimuli were univalent, i.e. one of the two relevant colours in an irrelevant shape, or one of the two shapes of an irrelevant colour. The background cue was presented for a long duration (3000ms) before the stimulus appeared, to allow longer for task preparation. Predictable runs of four repeat trials were followed by a single switch trial, thus only 20% of trials required a switch between task types and these switches were predictable.

Control Task (CT).

In order to control for the demands of each task whilst minimizing the demands on attention switching, control task conditions followed the same procedure as difficult switching conditions, but presented only one of the two task types and therefore no switches.

Control Difficulty (CD).

Performance costs are associated with task switching in typically developing individuals (Allport, Styles & Hsieh, 1994; Rogers & Monsell, 1995). Thus, when aiming to investigate the effects of a specific demand on attention switching (rather than just the effects of increased cognitive load), it was necessary to control for the increased difficulty associated with switching. To control for task difficulty, an equivalent procedure to the difficult switching condition was used, except that all stimuli were the same irrelevant picture (a closed box). Participants were told that as they could not see inside the box, they must guess the correct response. A random 33% of responses were followed by correct feedback, whilst 66% were treated as incorrect.

Stimuli and tasks.

Game 1 presented a daytime background cue to indicate the shape task and a night time cue to indicate the colour task. The stimuli were yellow or brown, crowns or boots, and participants were asked to collect treasure (yellow objects / crowns) by pressing the treasure chest button (Z) and throw rubbish away (brown objects / boots) by pressing the dustbin button (M). Games 2 and 3 were analogous, but used different background cues (beach/ river and countryside/ city), different stimuli (blue/ pink shells/ fish and red/ green buses/ cars) and instructions with a different background storyline.

Procedure

Sessions (single administration of one condition) lasted for five minutes and were usually administered in blocks (some at home and some at school) containing each of the four conditions. Five blocks over the course of 8 weeks were administered to RB;

six blocks (one with no easy switch session) over a period of eight weeks to AH; six blocks over four-weeks to AG; and seven blocks (one with additional difficult switch and control task sessions, and one with a single difficult switch session) over a period of four weeks to KT. Blocks were administered during between three and four visits to each participant, spaced between one and five weeks apart. Multiple blocks administered during the same visit took place between one and five hours apart. For each participant, blocks were counter-balanced for the order of the conditions and the type of game in each block.

Results

Performance during the switching challenges

Performance during the switching challenges was measured using reaction times (faster reaction times indicate better performance) and accuracy rates. In order to account for possible speed-accuracy trade-offs we calculated a composite performance score by dividing the mean reaction time in each session by the proportion of correct responses (e.g. see Lavi et al., 2007), lower composite scores therefore indicate better performance. Figure 1 shows the mean composite scores for each participant in each switching challenge condition. Wilcoxon Signed Ranks tests were conducted to compare each individual's performance across the conditions. Analysis of performance in this way allowed us to test the experimental integrity of the switching challenges.

[Figure 1 about here]

Critically, the control conditions were designed to place equivalent general cognitive demands on participants to the switching conditions, without involving any attention switching. In support of this aspect of experimental integrity, there was no significant difference in performance between Easy Switching and Control Task conditions shown by any of the participants (RB: p= .500; AH: p= .500; AG: p= .345; KT: p= .465). Also supporting experimental integrity, RB showed no significant difference in performance between Difficult Switching and Control Difficulty conditions, and the other three participants showed significantly or bordering significantly better performance in the Difficult Switching relative to the Control Difficulty conditions (see Figure 1).

With the Easy Switching condition we aimed to manipulate the Difficult Switching challenge using the parameters that have been shown to affect switching performance in the general population, in order to reduce the demands on switching. These manipulations were successful and all participants showed better performance in the Easy Switching condition relative to the Difficult Switching condition (see Figure 1).

Behavioural results during the switching challenges

Figure 2 shows the percentage of time when temper outburst related behaviours was shown by participants RB, AG and KT across consecutive sessions. AH however did not show any temper outburst related behaviours during these sessions. Effect size for the differences in behaviour shown by each participant across different conditions was calculated using Cliff's *d* statistic, which measures the extent to which one sample distribution tends to lie above another. It has been demonstrated that this statistic is robust when employed with non-normal distributions, even when variances differ (Cliff, 1993). A *d* value of 1.0 indicates that there is no overlap between the two sampling distributions. Cliff's *d* is directly related to the Wilcoxon-Mann-Whitney *U* statistic (Cliff, 1993), so in addition to effect size, the comparisons between difficult switching, easy switching and control conditions were also tested for significance using *U* values.

[Figure 2 about here]

In difficult switching sessions RB, AG and KT showed more frequent temper outburst related behaviours than in control sessions. This difference in behaviour was associated with large effect sizes (*d*=(RB) 0.73; (AG) 0.86; (KT) 0.58) and was significant (RB (U=8.5, p= .041); AG (U=5.0, p= .004); KT (U=22.0, p= .03). In easy switching sessions RB, AG and KT tended to show more frequent temper outburst related behaviours than in control sessions, but these differences were only associated with small to medium effect sizes (*d*=(RB) 0.38; (AG) 0.40; (KT) 0.46) and were not significant RB (U=16.5, p= .292), AG (U=21.5, p= .174) and KT (U=21.0, p= .114). However, the frequency of temper outburst related behaviours shown in difficult switching sessions was not significantly different from that shown in easy switching sessions in RB (U=5.5, p= .142), AG (U=6.0, p= .055) or KT (U=13.0, p= .156). Therefore, having to switch was associated with more temper outburst related behaviours than not having to switch, but when the demand on switching was reduced (in the easy switching condition), so was the frequency of temper outburst related behaviours.

Discussion

In support of the hypotheses, three participants showed more frequent temper outburst related behaviours during switching conditions compared to control conditions. Also, these three participants showed less frequent temper outburst related behaviours in the easy switching condition compared to the difficult switching condition. Thus, experimental control was demonstrated over temper outburst related behaviours simply by manipulating the level of demands placed on the specific cognitive capacity of attention switching. This provides some evidence that a deficit in attention switching can underpin these difficult behaviours, although it remains necessary to investigate how demands on attention switching may occur in every day life.

Looking at the occurrence of temper outburst related behaviours across different sessions of the switching conditions, it is apparent that there were occasions when the frequency of behaviours in the switching conditions was not higher than in the corresponding control conditions. Therefore, it appears that at some times participants were less likely to show potentially difficult behaviours following a demand on attention switching. A possible explanation for this is that levels of engagement with the task may have differed across sessions. Indeed, it has been demonstrated that in typically developing children and children with ADHD, differing classroom contexts can affect on-task behaviour (Lauth, Heubeck & Mackowiak, 2006). If a participant showed little engagement with the task during a particular session, one would expect the cognitive demand of that session to be reduced, leading to a lower occurrence of the behaviours that follow that demand. However, variability of behaviour across different switching sessions could also reflect context dependent differences in the ability to cope with the proposed arousal caused by the demand on switching, or in the level of arousal produced. Future research presenting switching challenges in a variety of different contexts would help to address this issue.

It is interesting to note that the participant (AH) who did not ask any questions or show any temper outburst related behaviours, was also the participant who showed the best performance in the switching conditions. It may be therefore, that the switching tasks did not place a high enough demand on AH's attention switching capacity to trigger any potentially difficult behaviours.

We calculated the composite switching challenge performance scores using a method that has been used previously in the literature. In this way, we made the assumption that speed and accuracy were equal indicators of task difficulty and overall cognitive demands, which is not necessarily the case. In light of this it must be noted that in the Control Difficulty condition reaction times were generally faster than in the other conditions, so the main contributor to decreased performance in this condition was the (experimentally manipulated) low accuracy rate. However, the combination of the four conditions that place differing demands on attention switching and general cognitive resources makes it possible to separate the effects of increasing switching demand from those of increasing general cognitive demands.

EXPERIMENT 2

The objective of this experiment was to investigate the environmental mechanism (unexpected change to routines or expectations) that it is hypothesised can lead to a high demand on attention switching cognitive resources, and thus trigger temper outbursts and repetitive questions. Therefore, Experiment 2 investigated the effects of a specific environmental antecedent (unexpected change to routines or expectations) on temper outbursts and repetitive questions.

To achieve this, two experimental conditions were administered via tabletop games in which either, routines and expectations were established and followed as expected, or routines and expectations were unexpectedly changed. It was hypothesised that when exposed to these conditions, participants would show more temper outburst related behaviours when unexpected changes were imposed on routines or expectations, relative to when routines or expectations were followed as expected.

Methods

Measures

Change Challenges

Two types of analogue condition were presented via a variety of different tabletop games in order to avoid practise effects and maintain a high level of unpredictability during change sessions. These tabletop games are described in Table 3. Additional to the experimental manipulations described below, the winner of each game and the occurrence of any other potentially rewarding events were controlled in order to ensure that experimental manipulations were not confounded by differential levels of access to preferred events.

[Table 2 about here]

No-change condition.

During this condition a game was played which the child was already accustomed to playing and the child's normal routines and expectations around this game were followed. Alternatively, a new game was introduced to the child, expectations were made explicit and routines were established, and these were followed during play. For example, the *no-change* condition of the school counting game (activity 1) with RB, involved following the routines that the total number of sweets must be added following rolls of two dice and that following the roll of the second die within each round, the sweets must be placed on the picture that does not yet have any sweets.

Unexpected Change condition.

During this condition unexpected changes were imposed to the routines or expectations that existed within the game being played. Each game was associated with a number of routines or expectations and hence a number of possible changes could be applied. Changes were imposed intermittently, so play continued as expected for a short period (i.e. 10-20 seconds) before a change was imposed. In order to minimize the predictability of the changes, each type of change was followed by a change of a different type. For example, the *unexpected change* condition of the school counting game with RB could have involved adding the total number of sweets after more or less than two rolls of the dice, or placing the sweets corresponding to the roll of the second dice on a picture on which sweets had already been placed.

Procedure

Experimental sessions were five minutes long except fourteen two-minute sessions administered to AH. A total of 20 sessions were administered to RB; at home and at school over nine weeks. 36 sessions were administered to AH over nine weeks; all took place at school apart from the last ten (home). AG took part in 21 sessions, the first five at school and the remainder at home, over four weeks. KT took part in 24 sessions over five weeks; all sessions were administered at school except for the final eight sessions (home). Critically, despite the differing contexts of the Unexpected Change and No-change conditions the only difference between each No-change and corresponding Unexpected Change condition was that changes were imposed on the established routines and/or expectations.

Results

Figure 3 shows the percentage of time during which RB, AH, AG and KT showed temper outburst related behaviours across sessions. All participants showed higher frequencies of temper outburst related behaviours in the Unexpected Change condition compared to the No-change condition. This difference in behaviour was generally associated with large effect sizes (d=(RB) 0.98; (AH) 0.32 (AG) 0.84; (KT) 0.83). We used the Wilcoxon Signed Ranks test (WSR: comparing each unexpected change session with the preceding no change session) to test these effects for significance (e.g.

Cliff, 1993). This test allowed us to control for the variability in behaviour that was associated with the changing activities. The difference in frequency of temper outburst related behaviours between change and no change conditions was significant for all participants (RB: p= .005; AH: p= .028; AG: p= .005; KT: p= .003).

[Figure 3 about here]

RB and KT showed a trend of decreasing frequencies (following an initial increase in RB) of temper outburst related behaviours across consecutive Unexpected Change conditions only. In AH temper outburst related behaviours only occurred during unexpected change conditions, but also only during two of the activities (snap and scrabble). These two activities were the only activities during which any individuals other than the participant and the researcher (a classroom assistant or the participant's brother) were present.

Discussion

In support of our hypotheses, all participants showed increased temper outburst related behaviours in the Unexpected Change condition relative to the no-change condition. Experimental control was therefore demonstrated over temper outburst related behaviours through manipulation of the level of environmental predictability, in the same way that control over these behaviours was exerted through manipulation of the cognitive demands on attention switching. This provides additional evidence that a deficit in attention switching can underpin difficult behaviours, by identifying an environmental mechanism (changes to routines or expectations) that can place demands on attention switching in real life. It remains necessary however, to show that changes to routines or expectations that occur naturally within a day to day context, can trigger the same cluster of behaviours.

It is interesting that the only two activities that triggered temper outburst related behaviours in AH, in Unexpected Change sessions only, were also the only two activities presented to AH in the presence of an additional familiar person other than the researcher. It was also AH who did not demonstrate any temper outburst related behaviours during Experiment 1. The fact that the temper outburst related behaviours were limited to Unexpected Change sessions suggests that the change (and, we would hypothesise, the demand on attention switching) was a necessary factor in triggering these behaviours, however in this case it was not sufficient. Perhaps due to AH's relatively high ability in attention switching, in most of the situations presented, she was able cope with the imposed demand on switching, but the presence of another familiar person either added to the level of arousal experienced due to the switching demand, or increased the likelihood that a behaviour would be shown following that arousal. Anecdotally, the temper outburst related behaviour tended to occur when the additional familiar person emphasised the fact that a change had occurred, perhaps making it more salient. Alternatively, the presence of a familiar person may have acted as a discriminate stimulus; cuing the availability of predictability (i.e. AH might expect people that she knows well to provide predictability as they have done in the past).

The generally decreasing trends in behaviour frequencies shown by RB and KT across Unexpected Change but not across No-change conditions suggest that there is an effect of repeated exposure to changes, not just within the same activity, but also within similar but explicitly unrelated activities carried out by the same researcher. This raises the issue that these participants may have been learning about the researcher's behaviour and that changes often occurred with this person. Perhaps therefore, the 'unexpected' changes were becoming more predictable as the routines and expectations associated with doing any of these kinds of activities with the researcher were being progressively weakened.

EXPERIMENT 3

The objective of this experiment was to extend the social validity of Experiment 2 by investigating if temper outburst related behaviours could be triggered in the natural environment by imposing changes to normal routines or expectations.

In this experiment it would not be possible to exert experimental control over the behaviours, because necessity to observe behaviour in the natural environment forced the environment to remain uncontrolled. However, natural observation of the participants would have been unlikely to include enough incidences of the behaviours and antecedents of interest to produce sufficient data for analysis. Therefore, a method was used that was adapted from Structured Descriptive Assessment methods (e.g. Anderson & Long, 2002), which allow potential antecedents to be programmed into the observation period. A number of routines and activities involving expectations were identified for each participant. Each of these routines/activities was observed at least once in the No-change condition when no antecedents were programmed, and at least once in the Unexpected Change condition when a change was imposed to the expected routine or activity. It was hypothesised that more temper outburst related behaviours would occur in the Unexpected Change condition relative to the No-change condition.

Methods

Measures and Procedure

Three routines or activities involving expectations were identified for each participant and were observed on between two and four different occasions when a) routines and activities occurred normally as expected and b) an unexpected change was imposed to the routine or expectation. Further details about the routines and activities observed and the procedure applied are shown in Table 3. Critically, although the environment was uncontrolled, across the observation period unexpected changes only ever occurred during Unexpected Change sessions and the presence or absence of unexpected changes was the only factor that varied consistently across the two conditions.

[Table 3 about here]

Results

Figure 3 shows the percentage of time in which RB, AH, AG and KT showed temper outburst related behaviours across observation sessions. It can be seen that all participants showed greatly increased frequencies of temper outburst related behaviours in unexpected change observation sessions, compared to no change sessions. These frequency differences were associated with very large effect sizes (*d*=(RB) 0.88; (AH)

1.0 (AG) 1.0; (KT) 1.0), and although tests for significance must be treated with caution due to the small number of data points, the effects were significant in all participants (RB (U=4.0, p= .003); AH (U=0.0, p= .007); KT (U=0.0, p= .036) except in AG where the effect bordered significance (U=0.0, p= .057).

[Figure 3 about here]

The participants who were observed crying (RB, AH and AG), only showed crying during unexpected change sessions. In all participants the extent of the behavioural response to unexpected changes was highly variable across the different contexts in which the change occurred.

Discussion

In support of the hypothesis, all participants showed increased temper outburst related behaviours during observation sessions in which unexpected changes were imposed to routines or expectations in the natural environment, compared to sessions during which routines and activities occurred as expected. Therefore, by manipulating the level of predictability in the natural environment, the same behaviours could be triggered as those that were experimentally controlled by placing cognitive demands on attention switching in Experiment 1 and by imposing unexpected changes on routines or expectations in Experiment 2. This completes the chain of associations between a cognitive antecedent to behaviour and an environmental mechanism (unexpected changes) that could lead to everyday life manifestations of that antecedent, therefore providing additional evidence that a deficit in attention switching can underpin repetitive questions and temper outbursts in PWS.

The variability of the degree of behavioural response to changes to routines and expectations in the natural environment depending on the context of those changes, suggests that additional environmental factors are important in determining how individuals respond to changes. One possibility is that different changes to routines or expectations may produce differing levels of cognitive demand on attention switching, perhaps for example due to expectations being weaker or stronger, which could result from different routines being more or less established. Another possibility is that other environmental factors affect how individuals are able to cope with any particular level of demand on attention switching. For example, we discussed in Experiment 2 how the presence of familiar people may have resulted in increased likelihood of temper outburst related behaviours being shown following changes by AH.

GENERAL DISCUSSION

In a series of three experiments we have demonstrated experimental control over repetitive questions and temper outburst related behaviours by manipulating a specific cognitive antecedent (demand on attention switching) and then by manipulating the level of predictability in an experimental environment (showing how a specific environmental mechanism can lead to this cognitive antecedent occurring in everyday life). Additionally, we were able to demonstrate that the repetitive questions and temper outburst related behaviours could be triggered in a natural environment by introducing unexpected changes to routines or expectations. We have therefore provided support for a directional association between a cognitive deficit in attention switching, a resistance to change, repetitive questions and temper outbursts in four[4] individuals with PWS.

In Experiment 1, it was demonstrated that it was possible to reduce the demands on attention switching in four individuals with PWS using factors (increased: preparation time, predictability and discriminable properties of the stimuli) that can decrease the switching demands in the normal population. The reduced demand on switching was associated with lower frequencies of potentially difficult behaviours shown by participants and thus it can be seen how these factors could be translated into potential behavioural coping strategies. However, our results do not inform on the individual level of effectiveness of each factor in reducing the demand on attention switching and therefore it would be useful in future research to attempt to separate the different factors to provide additional information for the design of effective coping packages.

The hypotheses that we have previously proposed (withheld for blind review) include the suggestion that the deficit in attention switching in PWS is linked to repetitive questions and temper outbursts via a state of high arousal that occurs when demands are placed on switching capacity. Although this hypothesis could not be rigorously tested in the present study we were able to obtain pilot data on heart rate during the switching challenges using a portable heart rate monitor. Given the close equivalence of movement during the different switching challenge conditions, heart rate measures could be taken as a reasonable indicator of physiological arousal (Berntson et al., 1997;

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[4] In one of the four participants (AH), the demand on attention switching through cognitive challenge did not trigger temper outburst related behaviours or repetitive questions. Thus, in AH a relationship was demonstrated between resistance to change, temper outbursts and repetitive questions; however the association with the deficit in attention switching remains hypothetical.

Keysor, Mazzocco, McLeod & Hoehn-Saric, 2001; Pumprla, Howorka, Groves, Chester & Nolan, 2002; Roberts, Mazzocco, Murphy & Hoehn-Saric, 2008). Analysis of pilot data suggested decreased heart rate variability in switching relative to control conditions, which can be taken as an indication of increased physiological arousal (Delaney & Brodie, 2000; Hofmann, Moskovitch & Kim, 2006; McCann et al., 1993; Sgoifo, Stilli, de Boer, Koolhaas, & Musso, 1998). Future research investigating physiological arousal associated with demands on attention switching in individuals with PWS could therefore provide important insights into the mechanisms that may act to cause an association between a demand on switching and behaviour.

In Experiment 2 it was discussed how decreasing trends in the frequency of participants' difficult behaviour during unexpected change analogue sessions suggest that participants' expectations were being weakened as they learnt to expect unpredictable behaviour from the researcher. Also discussed above is how AH may have learnt to expect predictable behaviour only from familiar people. In support of these accounts, it has been shown that young children consider an informant's perceived credibility (based on the informant's previous behaviour) when learning new object labels (e.g. Jaswal & Neely, 2006). This introduces an exciting prospect for potential intervention strategies that might aim to associate a discriminative stimulus (in this case a person, but potentially an object) with unpredictability.

The experiments have shown that a demand on attention switching can trigger the same temper outburst related behaviours that can also be triggered by changes in routines or expectations in these individuals with PWS. However, there is no evidence to suggest that these behaviours can only be caused by demands on attention switching via changes. One would predict that any event that placed a demand on attention switching could also cause such behaviours in these individuals, as could any event that produced a state of high arousal. Important to note here is that full-blown temper outbursts as described by parents and carers were only ever observed during the natural environment observation sessions. Therefore, the possibility must be considered that the aversive state produced by a demand on attention switching can be incremented by stimuli and events occurring in the natural environment, and so a pure switching demand may not be sufficient to produce a full-blown temper outburst. The present results are therefore limited in only describing a single route between cognition and behaviour; further research into other routes and how additional factors may interact with the pathway described here, would be interesting.

The case study approach applied limits how far these results can be generalized to all individuals with PWS and how far the findings may be specific to individuals with the syndrome. However, the important result here is that it is possible for a specific cognitive deficit in attention switching to trigger temper outbursts and repetitive questions via a particular environmental mechanism (unexpected changes), in individuals with PWS. Previous research has shown that children with PWS show a deficit in attention switching (withheld for blind review) and a higher level of preference for routine, repetitive questions and temper outbursts relative to comparison groups (Holland et al., 2003; Moss et al., 2009). Thus it is possible that this pathway from cognition to behaviour could generalise to other individuals with PWS and show some degree of specificity for this population. However, attention switching deficits and resistance to change have been reported in other populations (e.g. boys with Fragile X syndrome: Moss et al., in press; Wilding, Cornish & Munir, 2002), therefore the pathway may not be entirely (if at all) specific to PWS. Future research using group designs should be able to address these issues.

One fundamentally important point which at first glance may appear as a limitation to the present results but in fact is the opposite, is the variability of contexts across different blocks of sessions in the three experiments. It could be argued that varying contexts led to a reduction in experimental control. However, the important point is that experimental conditions (switching/ Unexpected Change conditions) were always conducted in exactly the same context as the corresponding control conditions. Thus, the variability of context across different blocks of sessions added to the validity of the present results, showing that within each individual, affects of switching and unexpected changes on behaviour generalised across multiple contexts.

The methods applied here have thus provided a way of demonstrating proof of principle in relation to the mechanisms involved in a specific pathway between cognition and behaviour. It is hoped that this research will extend traditional approaches to behaviour change in people with intellectual disabilities by emphasizing the importance of a cognitive component.

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Table 1. Shows the general cognitive and behavioural characteristics of each of the participants.

Measure		**RB**	**AH**	**AG**	**KT**
Chronological age at testing (years)		13:0	13:0	11:4	16:0
Age equivalent years	WISC Similarities	6:2	7:6	9:6	6:2
	WISC Vocabulary	6:6	<6:2	6:2	<6:2
	WISC Block Design	<6:2	7:6	7:2	<6:2
	WISC Object Assembly	<6:2	6:6	6:2	<6:2
Simon Task Switch cost standard score (Z-score)		3.6	4.23	3.04	1.56
Repetitive questions RBQ item		More than once a day	More than once a day	More than once a day	More than once a day
Preference for routine RBQ item		More than once a day	Once a day	More than once a day	More than once a day
Preference for routine CRI item		Very much	Very much	Very much	Very much
Behaviours reported by carer to occur during a temper outburst		Ignoring, answering back, refusing, arguing, swaying of the head, angry facial expression, shouting, screaming, crying, salivating, stamping, red in the face, throwing things	Rigid posture, glaring, tapping the hands on surfaces, stamping feet, clenched fists, shaking hands, squealing/crying, disagreeing/saying no, shouting, salivating, red in the face, angry facial expression	Refusing requests, arguing, shouting, squealing, crying, salivating, red in the face	Questioning, rubbing hands, puzzled look, rigid posture, angry facial expression, red in the face, shouting, stamping, banging things, throwing herself down in chairs, ignoring, refusing requests

Table 2. Describes the tabletop games that were used to present blocks of change analogue sessions to each participant. Each activity allowed the administration of *unexpected change* and *no change condition* sessions. An average of four sessions (2 *unexpected change* and 2 *no-change*) of each activity were administered and all sessions lasted for five minutes except for those marked with an (*), which lasted for two minutes.

Participant: Activity	**Location**	**Description**
RB: 1	School classroom; class doing other activities.	**School counting game**. Rolling two dice alternately; assigning the corresponding number of sweets alternately to two A4 pictures of faces; adding the sweets on both faces. The participant had played this game approximately once a week for at least a year.
RB: 2	At the participant's home with the participant's mother observing.	**Pop-up Pirate game**. The participant and the researcher played a series of *Pop Up Pirate* games (made by Tomy). The participant had been accustomed to playing this game regularly with her family for at least a year.
RB: 3		**Card game**. The participant and the researcher took turns to count a number of cards out face down into a central pile, followed by a final card face up. The player showing the highest card received the cards from both piles. An alarm sounded after two minutes. This game was novel to the participant on the first occasion that they played it with the researcher.
AH : 1	In a quiet room at the participant's school.	**School sums**. Addition sums of two one-two digit numbers were given verbally and written for the participant to complete. The participant had been accustomed to this activity for approximately 6 months on a regular basis at school.
AH : 2		**School spelling**. Normal procedure of school spelling practise was followed. The participant had been accustomed to this activity for approximately 6 months on a regular basis at school.
AH : 3	Quiet room at school; observed by classroom assistant.	**Snap card game**. The participant and the researcher took turns to place a card in a central pile. When 2 consecutive cards were played that were the same as each other on some dimension, the first person to say snap received all of the cards underneath. The participant had practised this game at school over the preceding two weeks.
AH : 4 *	Classroom at school; class engaged in other activities.	**Counter game**. The participant and the researcher took turns to roll a dice (red / blue sides) and pick a card (red/ blue underside) from one of two piles. The player then received a counter for each of the colors that they had drawn. After two minutes an alarm sounder and the player with the most red counters won. The participant had played this game for approximately 30 minutes, two months before.
AH : 5 *		**Card game.** *As RB, activity 3*
AH: 6	Participant's home with her brother.	**Scrabble.** The participant, the researcher and the participant's brother played a game of *Junior Scrabble* (made by Mattel). The participant had been accustomed to playing this game with her family on a regular basis for a number of years.
AG : 1	Quiet room at school.	**Maths worksheet.** The participant was asked to complete a worksheet with the help of the researcher. This particular worksheet was novel to the participant, but she had been accustomed to completing similar worksheets in class for at least a year.
AG : 2	At the participant's home.	**Snap card game**. *As AH, activity 3 except that AG had been accustomed to playing this game with her family for a number of years.*
AG : 3		**Counter game**. *As AH, activity 4.* Except that this game was novel to the participant at the time of testing.
AG: 4		**Card game.** *As RB, activity 3*
KT : 1	In a quiet room at the participant's school.	**Connect-four game.** The participant and the researcher played a series of games of *Connect Four* (published by Milton Bradley, 1974). The participant had been accustomed to playing this game with classmates on a regular basis for at least one year.
KT : 2		**Counter game**. *As AH, activity 4.*
KT : 3		**Shopping game.** Shopping board game where the researcher & participant took turns to move between shops by rolling a die & purchase items using plastic money. The participant had played this game with classmates on an occasional basis for at least a month.
KT : 4		**Card game.** *As RB, activity 3*
KT : 5	At the participant's home.	**Memory game.** The participant and researcher took turns to turn over two of cards (from a deck laid out face down in a large rectangle) and kept the cards only if they showed the same number or picture. The winner was the player with most cards at when an alarm sounded after two minutes. The participant had played this game occasionally with family members over at least one year.
KT: 6		**Counter game**. *As AH, activity 4.*

Table 3. Describes the routines and activities in which participants were observed in their natural environment. The changes imposed to routines or expectations within the unexpected change condition sessions are described; in no-change sessions no change was imposed.

Participant: activity: number of sessions	**Date**	**Description**	**Routine / expectation**	**Unexpected change**	**Length of sessions** (minutes)
RB: 1: (9)	11-June (3); 2-July (2); 16-July (4)	Travel to and from swimming baths (half a mile).	Travel to baths in a wheelchair, walk back with a friend. This routine had been established on a weekly basis for the previous 10 months.	Walking when on the way to the swimming baths; walking back from the baths without a friend	5-30
RB: 2: (3)	11-June; 2-July; 16-July	Eating lunch at school.	Lunch includes three small pieces of fruit (e.g. two mandarins and a plum). This routine had been established for at least 2 years previously.	One large apple of equivalent size to the three pieces of fruit was included instead of the normal fruit in the lunch.	30-40
RB: 3: (4)	11-June; 2, 16-July; 10-August	Expectations about activities to be carried out in class or at home.	Either an expectation was made explicit about the activity to be carried out later the same day or no expectation was made explicit.	Specific expectations were changed at the last minute; one change resulted in not being able to play with the researcher as expected, the other involved being asked to play with the researcher for longer than expected.	18-46
AH: 1: (3)	12-June; 6, 18-July	Dinner money monitor.	To take the class' dinner money form the classroom to the office every morning (established 2 weeks prior to the start of testing).	Another member of the class was asked to take the dinner money.	8-10
AH: 2: (4)	12-June; 06, 18-July; 9-August	Eating lunch at school or home.	Expect to have either a packed lunch or hot lunch at school depending on the day, or expecting to have the meal agreed on in the morning for lunch at home.	Given a packed lunch when expecting a hot lunch at school. Given a different (but preferred) meal for lunch at home to what was previously agreed.	5-12
AH: 3 (2)	12-June; 18-July	Expectations about physiotherapist appointment.	Given specific expectation that would have an appointment at 2.00pm.	At 2.00pm told by teacher that there was not enough time to go to the appointment.	10, 21
AG: 1: (3)	13-June 5, 12-July	Expectations around eating lunch at school or home.	Expecting to eat a lunch prepared by the AG's nanny.	Told that she would have to eat something different at lunch time.	14
AG: 2: (2)	5, 12-July	Expectations around timing of activities.	Expecting to participate in games with the researcher at certain times.	Asked to participate in the games with the researcher at other times.	7, 20
AG: 3: (2)	5, 12-July	Morning cereal routine.	Morning cereal is prepared the morning before and kept on the work surface in the kitchen overnight.	AG was told by her nanny that the cereal must be moved into the cupboard because of the flying ants in the kitchen.	5-6
KT: 1: (3)	29-June 9, 13-July	Expectations about the classroom assistant.	KT had been in the same class as a particular classroom assistant for at least 1 year.	A different classroom assistant was present in the morning class with KT.	12
KT: 2: (2)	29-June 9-July	Class writing activity.	Writing exercise is copied from white board directly onto a computer document.	KT was asked to write the exercise on paper before typing it up.	29-30
KT: 3: (3)	29-June 9, 13-July	Break time expectations.	KT usually goes downstairs at break time to play with her friends.	Change of an explicit expectation that break time would be different from normal (changed back to normal).	18-19

FIGURE CAPTIONS

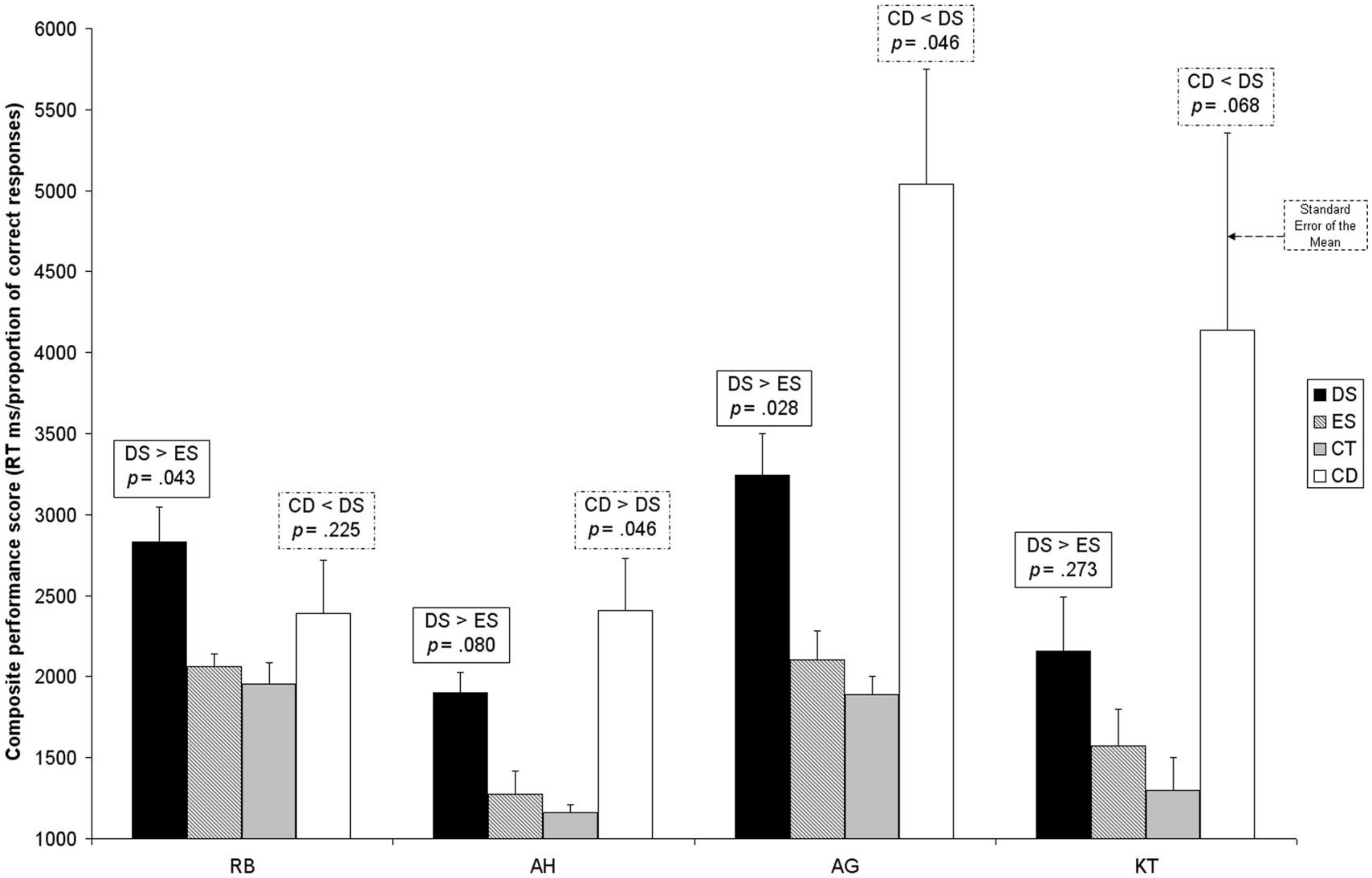
Figure 1. Shows mean performance across the four switching challenge conditions (DS, ES, CT & CD) for each participant in terms of composite speed accuracy score (RT/proportion of correct responses). *P*-values are shown for comparisons between DS and ES and between DS and CD conditions using Wilcoxon Signed Ranks tests.

Figure 2. Shows the percentage of time in each of the five-minute switching challenge sessions, when temper outburst related behaviours were shown by RB, AG and KT. No participants showed crying or ignoring in these sessions.

Figure 3. Shows the percentage of time in each of the change analogue sessions, when temper outburst related behaviours were shown by RB, AH, AG and KT.

Figure 4. Shows the percentage of time during each observation session when temper outburst related behaviours were shown by RB, AH, AG and KT. Looking at the researcher was not coded in these sessions as it was highly dependent on the particular activity being observed.

Figure 1



Composite performance score (RT ms/proportion of correct responses)
6000
5500
5000
4500
4000
3500
3000
2500
2000
1500
1000
RB
AH
AG
KT
DS > ES
p = .043
CD < DS
p = .225
DS > ES
p = .080
CD > DS
p = .046
DS > ES
p = .028
CD < DS
p = .046
DS > ES
p = .273
CD < DS
p = .068
Standard Error of the Mean
DS
ES
CT
CD

Figure 2

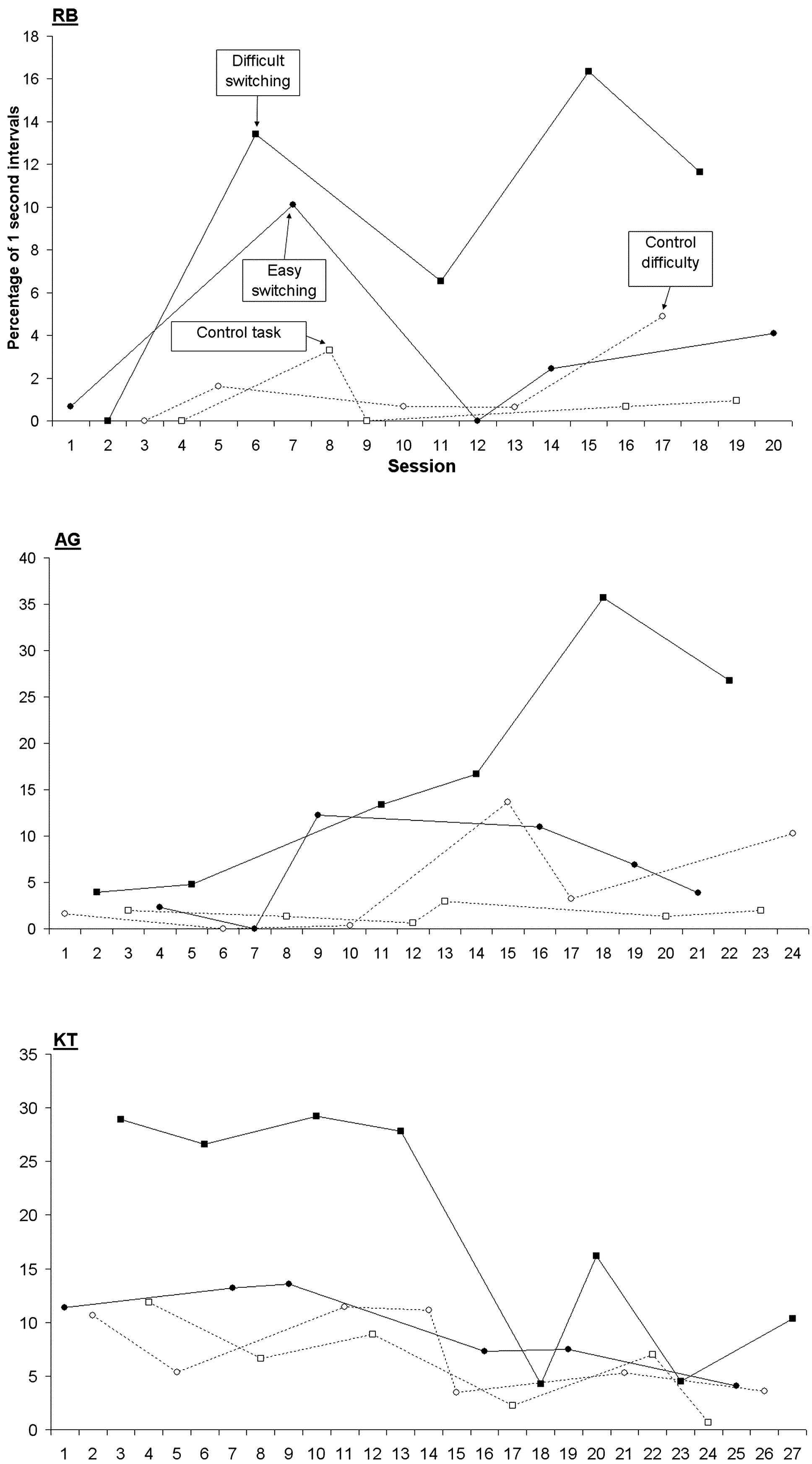

RB
Difficult switching
Easy switching
Control task
Control difficulty
Percentage of 1 second intervals
Session
AG
KT

Figure 3

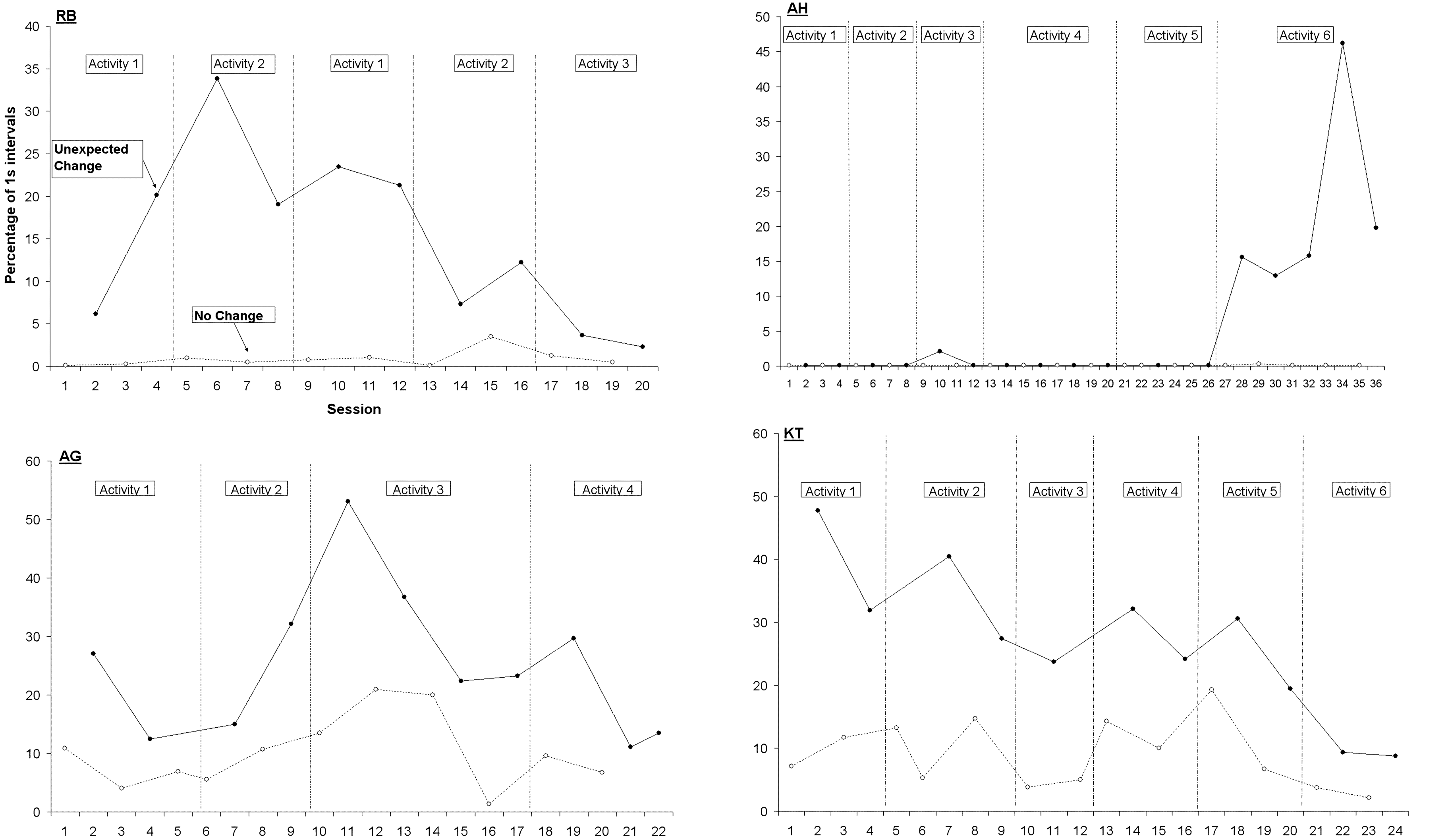
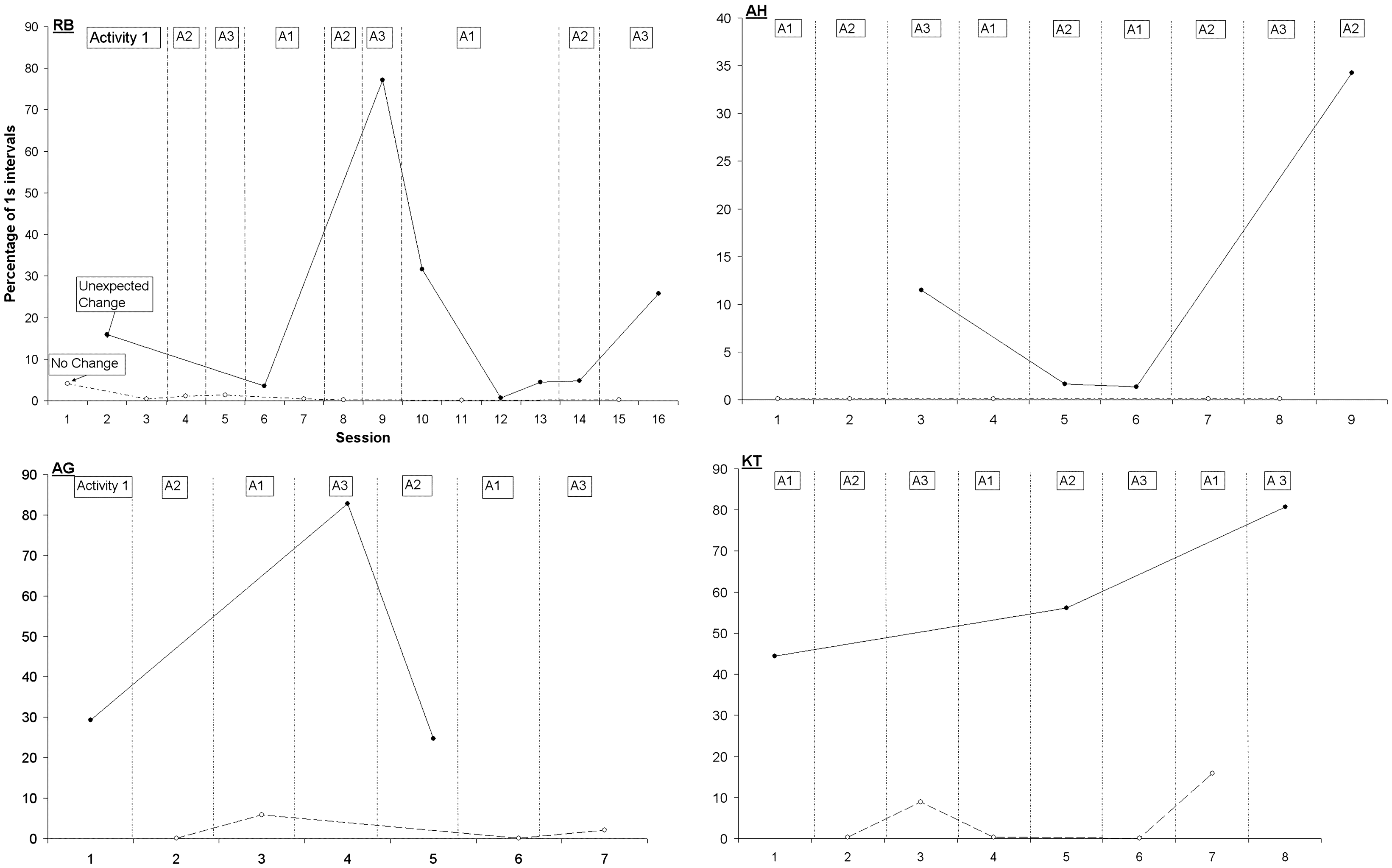

RB
Percentage of 1s intervals
Session
Activity 1
Activity 2
Activity 1
Activity 2
Activity 3
Unexpected Change
No Change
AH
Activity 1
Activity 2
Activity 3
Activity 4
Activity 5
Activity 6
AG
Activity 1
Activity 2
Activity 3
Activity 4
KT
Activity 1
Activity 2
Activity 3
Activity 4
Activity 5
Activity 6

Figure 4


RB
Activity 1
A2
A3
A1
A2
A3
A1
A2
A3
Unexpected Change
No Change
Percentage of 1s intervals
Session
AH
A1
A2
A3
A1
A2
A1
A2
A3
A2
AG
Activity 1
A2
A1
A3
A2
A1
A3
KT
A1
A2
A3
A1
A2
A3
A1
A 3