Associations between repetitive questioning, resistance to change, temper outbursts and anxiety in Prader-Willi and Fragile-X syndromes

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Abstract

Background

The behavioural phenotypes of Prader-Willi (PWS) and Fragile-X (FraX) syndromes both comprise repetitive behaviours with differences between the profiles. In this study we investigated the context and antecedents to the repetitive behaviours and the association with other behavioural phenotypic characteristics in order to generate testable hypotheses regarding the cause of the behaviours.

Method

The parents or carers of 46 children with PWS (mean age 14:1 years; 20 males), and 33 boys with FraX (mean age 13:11 years) were interviewed about their children's repetitive behaviour in a semi-structured format.

Results

Children showed negative emotional behaviour (PWS: 87.0%; FraX: 79.4%) and repetitive questions (PWS: 78.3%; FraX: 73.5%) following changes in routine or expectations. Significantly more temper outbursts were reported to follow changes in children with PWS (89.1%) compared to boys with FraX (41.2%) (χ^2= 20.93; p< .001). Anxiety that was frequently associated with repetitive and self-injurious behaviours in boys with FraX, followed changes in significantly more boys with FraX (76.5%) compared to children with PWS (6.5%) (χ^2=43.19, p< .001).

Discussion

On the basis of these reports and extant literature, we hypothesise that decreases in predictability are aversive to children with PWS and FraX. We also hypothesise that these children have a propensity to show a syndrome related pattern of behaviour (temper outbursts in PWS and displays of anxiety in

FraX) when an event in the environment has this aversive property. We hypothesise that questions may be reinforcing to children in their own right by increasing the predictability of the environment. We outline how a specific cognitive deficit in the endophenotypes associated with both PWS and FraX could be investigated as a potential explanation for the hypothesised aversive properties of decreased predictability.

Introduction

Prader-Willi syndrome (PWS) is a genetic disorder associated with mild to moderate intellectual disability. The syndrome is equally prevalent in males and females and evident in 1:25000 live births (Whittington, Clarke, Webb, Butler, Clarke & Boer, 2001). There are two main genetic causes of PWS: a paternal deletion within the chromosome 15 q11-q15 region (60-70%), and maternal uniparental disomy (UPD) of chromosome 15 (25-30%). Less than five percent of individuals show chromosomal translocations or mutations of the imprinting centre (Boer, Holland, Whittington, Butler, Webb & Clarke, 2002). The physical phenotype of PWS includes distinctive facial characteristics, hypotonia, hypogonadism, small hands and feet, short stature and hypopigmentation. Hyperphagia usually onsets in infants between the ages of 1 and 6 years, following a poor suckling reflex in infancy (Cassidy 1997).

A behavioural phenotype can be defined as "the heightened probability of, or likelihood that people with a given syndrome will exhibit certain behavioural sequelae relative to those without the syndrome." Dykens (p. 523, 1995). The behavioural phenotype of PWS comprises excessive eating, specific repetitive and self-injurious behaviours, temper outbursts, mood disturbance, excessive daytime sleepiness/ under activity and lying/stealing/ 'disobedience' (Dykens, Hodapp, Walsh & Nash, 1992; Clarke & Boer, 1996; Dykens & Kasari, 1997; Feurer, Dimitropoulos, Stone, Roof, Butler & Thompson, 1998; Einfield, Smith, Durvasula, Florio & Tonge, 1999; Richdale, Cotton & Hibbit, 1999; Symons, Butler, Sanders, Feurer & Thompson, 1999; Wigren & Heinmann, 2001; Dimitropoulos, Feurer, Butler & Thompson, 2001; Clarke, Boer, Whittington, Butler, Holland & Webb, 2002; Waltz & Benson, 2002; Webb, Whittington, Clarke, Boer, Butler & Holland, 2002; Holland, Whittington, Butler, Webb, Boer & Clarke., 2003; Wigren & Hansen, 2003; Holsen & Thompson, 2004; Oliver, Berg, Burbidge, Arron & Moss, in review; Arron, Oliver, Berg, Moss & Burbidge, in review; Moss,

Oliver, Arron, Burbidge & Berg, in review).

Fragile X syndrome (FraX) is the most common hereditary cause of intellectual disability, present in one in every four thousand males and one in every eight thousand females (Turner, Webb, Wake & Robinson, 1996). The mutation of a single gene (FMR-1) on the X chromosome causes FraX (Fu et al., 1991) by affecting the production of a specific protein (Siomi, Siomi, Nussbaum & Dreyfuss, 1993). There are gender differences in the FraX phenotype because of its X-linked nature (Loesch, Huggins & Hagermann, 2004). Males with full mutation FraX usually show a greater degree of intellectual disability (moderate to severe disability) than females (normal IQ to mild disability) (e.g. Alanay et al., 2007; Riddle et al., 1998). The physical phenotype associated with FraX includes characteristic facial features, macroorchidism, flat feet, hyper-extensible joints, soft skin, high arched palate, dental malocclusion (incorrect positioning of biting surfaces), single palmar crease, pectus excavatum (concave sternum) and spine curvature (Merenstein, Sobesky, Taylor, Riddle, Tran & Hagerman, 1996). The behavioural phenotype associated with FraX comprises language impairment, social anxiety, gaze aversion, and repetitive and self-injurious behaviour, and evidence suggests that there is an association between autism and FraX (Feinstein & Reiss, 1998; Hatton et al., 2006; Oliver et al., in review; Arron et al., in review, Moss et al., in review).

It has been suggested that the repetitive behaviour in PWS is obsessive-compulsive like (Dimitropoulos et al., 2001; Holsen & Thompson, 2004), but this behaviour can be distinguished from the repetitive behaviour shown in Obsessive Compulsive Disorder (Feurer et al., 1998; Dimitropoulos, Blackford, Waldon & Thompson, 2006). It has also been suggested that the repetitive behaviour in PWS may be developmentally appropriate. However Wigren & Hansen (2003) showed that the profile shown in PWS was distinct from that shown in typically developing children. The repetitive behaviour shown in FraX has been investigated in terms of its autistic-like nature, but evidence suggests that there are

differences between the profiles of repetitive behaviour shown in FraX and autism (Belser & Sudhalter, 2001; Baranek et al., 2005; Moss et al., in review).

In attempts to describe the aetiology of the repetitive behaviours in PWS and FraX, potential associations between these behaviours and other behavioural phenotypic characteristics have been investigated. Although skin picking shown in PWS is repetitive (e.g. Wigren & Heinmann, 2001), it is associated with a distinct pattern of variance to the other repetitive behaviours (Feurer et al., 1998; Holland et al., 2003), suggesting that it may have a different underlying cause. In a questionnaire and interview study, Holland et al. (2003) used factor analysis to demonstrate an association between repetitive behaviours and temper outbursts in PWS. In boys with FraX, Belser and Sudhalter (2001) argue that repetitive speech may result from the hyperarousal shown in the syndrome (Hessl et al., 2002). Symons, Clark, Hatton, Skinner & Bailey (2003) found some evidence of an association between self-injury and changes in routine in FraX. Although it remains to be seen exactly how repetitive behaviours in PWS and FraX fit into the broader behavioural phenotypes of each syndrome, it is clear that different individual classes of repetitive behaviour may show distinct aetiologies (Moss et al., in review).

In terms of individual classes of repetitive behaviour, the profile in PWS is characterised by an insistence on the sameness in daily routines, hoarding, repetitive phrases or questions, ordering, and excessive cleaning (Dimitropoulos et al., 2001; Wigren & Hansen, 2003; Clarke et al., 2002; Moss, 2005; Moss, et al., in review). The profile in FraX is characterised by repetitive speech, stereotypical movements, lining up objects, just right behaviour, tidying and preference for routines (Feinstein & Reiss, 1998; Moss, et al., in review). Comparing the repetitive behaviour profiles between individuals with PWS and FraX, both appear to show equally high preference for routine, but individuals with FraX show more stereotypical behaviour, repetitive speech, lining up objects and tidying. We aimed to

investigate the context of the specific profiles of repetitive behaviours associated with PWS and FraX.

Research in this area has been limited due to its heavy reliance on standardized questionnaire measures. Questionnaires not designed specifically for the population under investigation are unable to detect any behavioural aspects that may be particular to that population. Using questionnaire designs, possible associations between different behaviours can usually only be implied using statistical evaluation of relationships between items, and no evaluation is possible of the context of behaviours that may help to explain their occurrence. However, some questionnaires have been specifically designed to identify possible functions of particular behaviours. Two such measures are the Motivation Assessment Scale (MAS: Durrand & Crimmins, 1992) and the Questions about Behavioural Function scale (QABF: Matson & Vollmer, 1995). Studies investigating the reliability and validity of these measures (e.g. Duker & Sigafoos, 1998; Freeman, Walker & Kaufman, 2007; Hall, 2005; Nicholson, Constantinidi & Furniss, 2006; Paclawskyj, Matson, Rush, Smalls & Vollmer, 2001; Shogren & Rojahn, 2003) generally point towards reasonable reliability and validity but highlight important considerations such as the lack of complete correspondence with experimental functional analysis and reduced reliability for low frequency behaviours. Importantly for our purposes the MAS and QABF are limited to investigating specific potential functions for behaviours (attention, escape, tangible, sensory, physical, non-social) which would prevent us from investigating potential associations between different aspects of the behavioural phenotypes of PWS and FraX and would preclude the identification of idiosyncratic behaviour functions that might be specifically associated with PWS and FraX.

We used an informant report semi-structured interview that focussed on the behavioural and environmental context in which the repetitive behaviours in PWS and FraX occurred. Informant report interview methods have shown to be effective in providing rich data (e.g. Deb, Hare & Prior, 2007, on the onset of symptoms of early dementia in Down syndrome). O'Neill, Horner, Albin, Storey &

Sprague (1990) have published a detailed Functional Analysis Interview to assess the functions associated with challenging behaviours. The interview begins with a description of the behaviours of concern and proceeds through a number of specific questions aimed to identify the functions of each of these behaviours, for example by looking at the predicting events and consequences of the behaviours. Although this method is an important tool for clinical practise, to our knowledge there are no published studies reporting its reliability or validity. The semi-structured interview method we employed followed many of the important principals highlighted by O'Neil et al. for example prompting for antecedents and consequences associated with behaviours as well as asking for other contextual information that may reveal ecological events that could affect the behaviours. However, we chose to employ a less restrictive approach than that outlined by O'Neil et al. because we wanted to ensure that our interview method had the potential to reveal any specific associations between different aspects of the PWS and FraX behavioural phenotypes.

We aimed to provide data to support the development of specific testable hypotheses that could help to explain the presence of repetitive behaviours in the two syndromes and describe how the repetitive behaviours may be associated with other behavioural phenotypic characteristics.

Method

Participants

Participants were one or two parents or carers of 46 children with PWS and 33 boys with FraX. Children with PWS were 20 males and 26 females between 6:0 and 19:0 years, with a mean age of 14:1; *SD*: 3:1. Thirty children showed a paternal deletion of chromosome 15, nine showed maternal UPD, and seven had a clinical diagnosis only of PWS. All carers of children with PWS were recruited via the Prader-Willi Syndrome Association-UK. Boys with FraX were between the ages of 9:0 and

19:0 years, with a mean age of 14:0; *SD:* 2:4 and were recruited via the Fragile-X society. All showed a full mutation of the FMR1 gene. [1] The only inclusion criteria were that participants lived within four hours travelling time from the research base (in the centre of England), and that the children were aged between 6 and 19 years and had a diagnosis of either PWS or FraX. Subtests from the *Wechsler Intelligence Scales for Children* (Wechsler, Golombok and Rust, 1992) (Similarities, Vocabulary, Block Design, Object Assembly) showed that as would be expected from the degree of intellectual disability generally associated with each syndrome (discussed above), children with PWS performed at a higher cognitive level on all subtests relative to the boys with FraX (e.g. Vocabulary: PWS mean raw score=18.5; FraX mean raw score=10.3; $t(74)=4.78$, $p< .001$).

Measures

Semi-structured interview

A semi-structured informant report interview about children's repetitive behaviours was initiated with the question: "*I am interested in any behaviours that (Child A) might show that you would describe as repetitive. This could be doing the same thing more than once, saying the same thing more than once, liking things in a particular way or liking particular routines."*

[1] Reports of genetic classification are based on parent reports of previous genetic tests carried out at a hospital or genetic counselling service.

Questionnaire measures

Two informant report questionnaire measures were used in order to investigate the concurrent validity of behaviour codes identified from the semi-structured interview. The *Repetitive Behaviour Questionnaire (RBQ)* (Moss & Oliver, 2008) asks participants to rate the frequencies of nineteen observable, operationally defined repetitive behaviours on a scale of 0-4 (never, once a month, once a week, once a day, more than once a day). Previous studies have shown strong inter-rater reliability across individuals with heterogeneous causes of intellectual disability, high test- retest reliability and strong concurrent validity e.g. there is a strong association between pairs of scores referring to the same behaviour on the RBQ and the Repetitive Behavior subscale of the Autism Screening Questionnaire (Moss, et al., in review). The *Childhood Routines Inventory* (CRI) (Evans et al., 1997) is an informant report questionnaire that requires participants to rate the frequencies of nineteen observable behaviours on a scale of 1-5 (never, a little, sometimes, quite a lot or very much). The measure has previously been used with normative samples of children between 8 and 72 months (Evans et al., 1997), in children with Down syndrome (Evans & Grey, 2000), in people with autism, and in people with PWS (Greaves, Prince, Evans & Charman, 2006).

Procedure

The interviews were all carried out at participants' homes except for two that were carried out over the telephone, one that took place at the child's school and one that took place in a community centre. All interviews were recorded using an audio cassette recorder, except the two telephone interviews and two during which the participants did not wish to be recorded, which were summarised in note form by the researcher (KW).

Following the initial interview question, participants were left to talk without interruption for as long as they were able. When further input from the interviewer was necessary, participants were prompted for

the usual context of any behaviours that had been reported without context. For example, if the participant had talked about their child asking questions over and over again but had not said what the questions were usually about or when they were asked, then the interviewer would ask if the questions happened at any particular time or were normally about a particular topic. Once the possible antecedents, consequences and nature of all of the behaviours that were initially reported had been described, the interviewer reminded the participant of any other behaviours stated in the initial question that had not already been talked about. So if routines had not been mentioned, the interviewer would ask if the child liked to have particular routines. Any other behaviours that were reported were also prompted for their context if necessary. The interviewer occasionally asked direct questions if responses appeared to be implying something that had not been stated clearly and explicitly. For example if participants reported that their child often asked a question once and then asked the same question again a few minutes later even though they had been told the answer the first time, then the interviewer may have asked directly if the child knew the answers to the questions they asked. Interviews lasted between 30 minutes and one hour. Questionnaires were administered following the interview to ensure that the questionnaires did not lead participants' responses in the interview.

Development of the interview coding scheme

A single researcher (KW) who conducted all of the interviews listened to the tapes and operationally defined codes that described the nature of behaviours reported or antecedents or consequences that were associated with those behaviours. As we aimed to generate hypotheses relating to the repetitive behaviour shown in PWS and FraX, it would be outside the present objectives to report individual behavioural descriptions. Therefore, interview codes were only defined for behaviours or events that were reported by at least ten participants. Transcriptions were made of 124 clips taken from eighteen interviews (23%) in which one or more codes were identified. A second researcher independently classified each clip for the presence or absence of each defined code. Kappa values indicative of the

extent of agreement between inter-raters for each code were calculated based on the proportion of clips in which the researchers agreed or disagreed on the presence or absence of the code. In subsequent analysis of the interview codes, we were only interested in whether or not a particular code could ever be identified within each interview (codes could have been identified repeatedly throughout an interview, but were only counted on the first occasion). It was considered that the above method of reliability assessment would provide a stringent test as it ensured that raters were considering the same statement when they agreed that a particular code could be identified.

Operational definitions for the interview codes are shown in Table 1. In defining the interview codes, it was also necessary to define *changes* (change in routine, in what was expected, to plans, to what was done normally, to how things usually were, to what the child thought was going to happen or to what someone had told the child would happen), *temper outbursts* ('tantrum', 'temper reaction', 'blip', 'angry outburst' or two or more of: shouting, screaming, stamping feet, storming off, throwing things, crying, going red in the face, being physically aggressive or shaking occurring together) and *repetitive questions* (spoken or signed questions or statements interpreted as questions by carers that were repeated over and over or that occurred within a series of similar questions that would result in the same answer).

Table 1 about here

The level of prompt given by the interviewer that was necessary before the first report of each code in each interview was also recorded. Inter-rater reliability for the level of prompting was calculated based on a sample of 30 transcribed clips each demonstrating one of the three prompt levels. 'No prompt' (Kappa: .93) was assigned where a code was identified in a report that directly followed the initial question or followed a partial repeat of the initial question. 'Partial prompt' (Kappa: .84) was assigned when a code identifying a specific relationship between two behaviours or events was reported

following the mention of one of those behaviours or events by the interviewer, e.g. if the interviewer asked 'so are there any particular times when he asks questions?' and the participant responded 'if something changed in what we were supposed to be doing'- in this case *repetitive questions following changes* would have been partially prompted. Alternatively, 'what does he look like when he is anxious?'- 'he bites his hand' – in this case *self-injurious behaviour when anxious* would have been partially prompted. 'Full prompt' (Kappa: .92) was assigned when a code was identified in a response that followed a direct question by the interviewer. So in this case the code would have been identified by the interviewer in the question and the respondent would have either expanded on this or just provided a yes/no answer. Very good inter-rater reliability was therefore obtained for all of the above codes and prompt levels, the remainder of the interviews were therefore coded by the first researcher from the audio tapes. The percentage of times that each code was identified at each prompt level is shown in Table 1. It is apparent that most occurrences of most codes were either associated with no prompt or a partial prompt, suggesting that the potential bias from the interviewer's behaviour was successfully minimised.

Concurrent validity of the codes identified in the interviews was assessed using items from the RBQ and CRI. An interview code was considered to correspond with a questionnaire item when both reported the same behaviour e.g. the three stereotypical behaviour items on the RBQ corresponded to the 'Stereotypical behaviour' interview code. Additionally, the report of a particular interview code may have directly implied the presence of particular behaviours measured in the questionnaires. For example, 'Change followed by repetitive questions' implies that repetitive questions must have been present and so corresponds to the repetitive questioning item on the RBQ. It was hypothesized that where a participant reported a particular interview code, they would report a higher score on the corresponding questionnaire item than if they did not report that interview code. There were significant differences between the mean scores of participants who reported an interview code and those who did

not report that code on all corresponding questionnaire items except one, supporting the concurrent validity of the interview codes. The difference was not significant in RBQ preference for routine scores between participants who did and did not report that change was followed by negative emotional behaviour. However, these participants did differ significantly on the corresponding CRI preference for routine item, so it is possible that differences in the wording of these two questionnaire items (the RBQ item included the word 'insists' on having routines, whereas the CRI item only used 'prefers') may have led to the differing levels of agreement. Mann Whitney U and corresponding *p* values for these comparisons are shown in Table 1. It must be noted that not all of the interview codes could be tested for concurrent validity directly. For example, although the interview code 'Change followed by repetitive questions' showed a high level of agreement with the repetitive questioning item of the RBQ, this is not a direct test of the validity of the interview code that states specifically that *change* is followed by repetitive questions. However these strong agreements between the reports of the same behaviours in both the interview codes and the questionnaire measures do provide evidence towards the concurrent validity of the interview codes.

Results

Initial inspection of the interview codes reported in children with PWS revealed no evidence of differences between different genetic subtypes or type of diagnosis (clinical or genetic). Considering the small numbers in UPD and clinical PWS subgroups, we combined these subgroups into a single PWS group for further analysis. The percentage of participants with PWS and FraX that reported each interview code was calculated. Chi-square analyses were used to compare the proportion of participants reporting each interview code across the two syndrome groups; a Bonferroni correction was applied to correct for the sixteen separate Chi-square tests carried out and hence differences were considered significant only when $p < .003$. The percentages of participants reporting each interview

code and comparisons between the two syndrome groups are shown in Table 2.

Table 2 about here

Repetitive questions, repetitive speech or restricted conversation about the future was reported in 91.3% of children with PWS and 72.7% of boys with FraX. Repetitive questions were reported to occur following changes to routines or expectations in 78.3% of children with PWS and 75.8% of boys with FraX and most of these children were reported to know the answers to the questions that were asked.

Most children with PWS (87.0%) and boys with FraX (78.8%) were reported to show negative emotional behaviour following changes, although anger and temper outbursts (reported to usually follow the same course or include the same behaviours in 41.3% of children with PWS) following changes were reported more frequently in children with PWS than in boys with FraX (χ^2= 17.36, p< .001; χ^2=21.97, p< .001), and anxiety following changes was reported more frequently in boys with FraX (χ^2=43.19, p< .001). In both groups of children however, it was reported that temper outbursts could sometimes occur when the child was perceived as manipulating the situation or that these outbursts could be reinforced. More than half of the boys with FraX were also reported to show repetitive speech, self-injurious or stereotypical behaviour when anxious, which was not reported in children with PWS, and 39.4% of the boys with FraX were reported to show anxiety in social or highly stimulating contexts. It was reported by over 50% of carers of children with PWS and over 30% of carers of boys with FraX that children did not appear to have control over temper outbursts or overt displays of anxiety (i.e. involving repetitive/ self-injurious behaviour).

Rigid thinking or the expectation that things would be as they were previously was reported more

frequently in children with PWS (73.9%) than in boys with FraX (12.1%). About half of all participants reported on if and how advance warning of changes would affect the child's behaviour following the change. Examples of the various descriptions of this warning process included: the behavioural reaction to the change improved as the length of notice increased; notice was deliberately not given in order to avoid particular behaviours in the time leading up to the change; some notice was deliberately given, but 'not too much'; no notice was necessary.

Repetitive activities were frequently reported in both groups of children (PWS: 67.4; FraX: 87.9), but these were reported to take a variety of different forms and not to consistently occur in any particular context. Repetitive watching of videos stood out as the most common repetitive activity in boys with FraX (41.4% of the boys who were reported to show repetitive activities). Stereotypical behaviour was also reported in both groups of children (PWS: 28.3; FraX: 39.4), but the only consistently reported context of this behaviour was (as described above) in boys with FraX when they were anxious.

Discussion

Changes in routines or expectations caused negative emotional responses in both groups and repetitive questions, common in both groups, often appeared to function to increase predictability (questions were frequently about the future and increased in frequency following changes in routine). Anger and temper outbursts appeared to follow changes in routine or expectations in more children with PWS than boys with FraX, but anxiety (that was often associated with overt repetitive and self-injurious behaviours) followed changes more frequently in boys with FraX. Anxiety was also reported in social and highly stimulatory contexts in boys with FraX.

Preference for routine and repetitive questioning in individuals with PWS and FraX has been reported

previously (Clarke et al., 2002; Moss et al., in review), it would therefore appear that these behaviours are not specifically associated with one syndrome. Temper outbursts, reported frequently in individuals with PWS (e.g. Dimitropoulos et al., 2001; Waltz & Benson, 2002), have been linked to repetitive behaviour (Holland et al., 2003); supporting the relationship that we found between temper outbursts and a particular class of repetitive behaviour (preference for predictability). High levels of anxiety have been reported in FraX, particularly shown in social situations (Hall, DeBernardis & Reiss, 2006; Hessl et al., 2002), which supports the frequent reports of anxiety in the boys with FraX in the present study, including some reports of anxiety in social contexts. As temper outbursts followed changes in more children with PWS, and anxiety that included overt repetitive and self-injurious behaviour, followed changes in more boys with FraX, these behaviours appeared to be more syndrome-specific reactions to change.

The question therefore arises as to why repetitive questions, temper outbursts in PWS and displays of anxiety in FraX occur following changes to routines or expectations. We hypothesise that decreases in predictability (caused by a change) are aversive to children with PWS and FraX. A mutual reinforcement model would then predict that escape from a change would be reinforcing for the child's behaviour, meaning that any operant behaviour that resulted in the removal of the change would strengthen. The removal of a change by the carer would allow the carer to escape from the child's behaviour (aversive to the carer) and would therefore also strengthen. A process of shaping may then occur whereby a child's behaviours become more and more aversive to the carer due to patterns of alternating reinforcement and extinction. This type of mutual reinforcement framework has been described in the context of self-injurious behaviour (Oliver, 1993, 1995), but is unable to account for the differing profiles of behavioural reaction to change in the two syndrome groups: there must be other factors that cause a specific type of behaviour to develop in a certain population.

Additionally therefore, we hypothesise that children with PWS and FraX have a propensity to show a syndrome specific pattern of behaviour (temper outbursts in PWS and overt displays of anxiety in FraX) when the environment acts to produce an aversive state in that child. Specifically we would predict that changes to routines or expectations could trigger temper outbursts in children with PWS and displays of anxiety in FraX, but that these behaviours could also be triggered by other events that produce an aversive state in the children. In FraX, as discussed above, social situations appear to be associated with anxiety and physiological arousal (Hall et al., 2006; Hessl et al., 2002), which would be consistent with the suggestion that social situations are aversive to individuals with FraX. Environmental effects on apparent genetic predispositions for a particular behaviour have been reported for example in Angelman syndrome, with the laughing and smiling behaviour that is mediated by the social environment of the individual (Oliver, Demetriades & Hall, 2002; Oliver et al., 2007). Self-injurious behaviour in Rett syndrome was also shown to function both to escape and to obtain social interaction depending on the stage of an individual's development, because the Rett syndrome developmental profile causes social interaction to be reinforcing or punishing at different stages (Oliver, Murphy & Crayton, 1993; Murphy & Oliver 1987). Self-injurious behaviour in Cornelia de Lange, Smith Magenis and Lesch-Nyhan syndromes has also been shown to be influenced by levels of social contact (Arron et al., 2006; Taylor and Oliver, In press, Hall, Oliver and Murphy, 2001).

Partially in line with the reinforcement model described above, it is possible that questioning may have developed to escape or avoid the aversive state associated with changes. Repetitive questions could be reinforced by carers (who provide answers that increase the level of predictability). However, it is possible that questions may also evoke responses from a carer that would decrease the level of predictability (supported anecdotally by reports from some carers of a 'the right way' and a 'wrong way' of answering questions with either concrete facts or comments like 'I don't know' or 'maybe'). We therefore hypothesise that repetitive questions may directly increase predictability (e.g. by allowing

repetition of how events will happen) and hence be reinforcing to the children in their own right, regardless of how they are answered by a carer. This is supported by the high percentage of participants in both groups who reported that their children knew the answers to the questions being asked. Specifically from this we would predict that it would not be possible to extinguish repetitive questions based on the planned alteration of carers' responses. Additionally we would predict that if a child were prevented from asking questions, the aversion that the child would feel following a change would increase relative to if the child we able to ask questions.

Importantly, we note that changes in routine or expectations were not always reported to result in repetitive questions or temper outbursts/ displays of anxiety. Therefore, it appears that some changes are more aversive to children than others. This leads to the question of why changes are aversive to children with PWS and FraX. One possibility is that decreases in predictability place particular demands on the cognitive system of children with PWS and FraX due to a specific characteristic or set of characteristics associated with the endophenotypes of both syndromes. In order to explain why changes in routine or expectations are not always followed by temper outbursts or displays of anxiety, we suggest that certain characteristics of a change would affect the resulting level of cognitive demand for a child with PWS or FraX. This would also provide the potential to explain individual variation in behaviour; even if all individuals with a particular genotype show the same endophenotypic characteristics, different individuals may be able to cope effectively with different levels of cognitive demand. Anecdotally, some parents who reported very few problems associated with changes in routine, reported deliberately varying their child's environment to prevent routines from developing from an early age, which may have led to early acquisition of the cognitive skills necessary to deal with changes. Specifically from this suggestion, we would predict that children with PWS and FraX would share a specific cognitive deficit and that this deficit would be related to the resistance to change shown by children with both PWS and FraX. This hypothesis is directly testable and would provide a useful

starting point for investigation of the nature of the endophenotypes associated with PWS and FraX. However, it must be noted that this is only one of a number of possible interpretations of why changes may be aversive for children with PWS and FraX.

We have proposed a number of specific hypotheses that suggest important potential avenues for future research. We have made the distinction between syndrome specific and syndrome shared behaviours, but the actual level of specificity remains to be investigated. Although we have described temper outbursts as PWS-specific and overt displays of anxiety as FraX-specific, we only intend to suggest that each of these behaviours is more common in one syndrome compared to the other. Comparative studies with groups of people with intellectual disabilities of mixed aetiologies and other genetic syndromes would help to shed light on these issues.

The main limitations associated with the current study arise from its reliance on interview data and biases related to the interviewer's behaviour, and demand characteristics created by the interview environment. We choose to employ an open ended semi-structured interview instead of a closed ended interview approach such as that by O'Neill et al. (1990) or a questionnaire measure of behavioural function. Although it could be argued that these more structured approaches may have been associated with less biases related to the interviewer's behaviour, it must be noted that some of the important findings discussed may not have been uncovered using a more structured approach. For example preference for routine was reported by the majority of carers, following which it was important to find out what happened if children's routines changed and contextual information concerning any behaviours that may follow. This information would have been less likely to have been reported within a more structured approach.

As we highlighted above, the data that we present in support of the concurrent validity of the interview

codes do not test the validity of all of the codes directly, however the very specific nature of many of the codes means that it would be difficult to test their validity by comparison to previously published informant report measures. Therefore, it could be argued that behavioural observation would be a better test of the concurrent validity of the interview codes. However, it must be noted that strong discrepancies have been reported in the past between the observation of natural behaviour and informant based questionnaires of behavioural function (e.g. Hall, 2005) and that some of this at least may be due to changes in participants' behaviours that occur when individuals know that they are being observed. Additionally, validation of the present interview codes with behavioural observations would reduce one of the main advantages of the present method in that it is a very efficient way to generate new testable hypotheses. Indeed, future studies directly testing these hypotheses will provide important tests of the validity of the present method.

There are statistical limitations to the present study due to the descriptive nature of the data that were obtained. A further limitation arises from the assumption that fewer people reporting a behaviour in a certain population implies a lower frequency of the behaviour in that population. Strictly speaking, evidence of a lower frequency of a particular behaviour could only come from more people reporting that the behaviour does not occur. However, this type of study has important strengths due to the richness of the data, allowing the generation of testable hypotheses relating to connections between different behaviours and contexts. This may not be possible with data of a more quantitative nature, or would require more extensive research and analysis in order to reach the same hypotheses. Another important advantage arising from the semi-structured nature of the interviews is the capability of this technique to identify areas that are of most concern to parents and carers, as these areas are likely to be emphasised to a greater degree by participants. This means that future research is more likely to lead to positive results for the people involved.

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Table 1. Shows Kappa values relating to the level of agreement between inter-raters for each interview code. Also shown is the percentage of each level of interviewer prompting that preceded each code. Mann-Whitney U and *p* values are shown for the comparison of the mean scores of individuals who did or did not report each interview code, on the corresponding RBQ and CRI items. As expected, in all comparisons mean RBQ/ CRI item scores were higher in individuals who reported the corresponding interview code than in individuals who did not report that code.

Behaviour code *Operational definition* [2]	**Inter-rater reliability: Kappa**	**Percentage of the reports identified at each prompt level**			**Corresponding item(s) in RBQ or CRI**	**Mann Whitney U and *p* values for the comparison of scores on RBQ/ CRI items between individuals who did or did not report each code**
		None	**Partial**	**Full**		
Expect things to be as previously/ rigid thinking *Child thinks/ expects/ wants something to happen as it did before, or has 'rigid', 'not flexible' or 'black and white' thoughts, or has ideas that are difficult to change.*	0.78	60.53	10.53	28.95	CRI: 'Prefers to have things done in a particular order or in a certain way'	*U*=558.50, *p*= .024
Change followed by negative emotional behaviour *Child described as 'upset', 'not liking it', crying, 'tearful' or 'unhappy' after change(s) had occurred.*	0.92	52.24	46.27	1.49	RBQ: 'Preference for routine: insists on having the same household, school or work schedule every day' (1) CRI: 'Prefer the same household schedule or routine every day' (2)	(1) *U*=330.0, *p*= .107 (2) *U*=233.5, *p*= .002
Change followed by anger/ aggression *Child described as 'angry', 'aggressive', 'violent' or 'cross' following change(s).*	1.0	51.22	43.90	4.88	RBQ: (1) CRI: (2)	(1) *U*=583.5, *p*= .032 (2) *U*=606.0, *p*= .030
Change followed by temper outburst *A temper outburst was reported to occur following change(s).*	0.93	50.91	52.73	0.00	RBQ: (1) CRI: (2)	(1) *U*=468.0, *p*= .015 (2) *U*=463.5, *p*= .008
Change followed by anxiety *Children were described as being 'anxious', 'stressed', 'nervous' or 'mithered' after change(s).*	0.91	55.17	34.48	10.34		
Change followed by repetitive questions *Repetitive questions followed change(s).*	0.83	44.26	44.26	11.48	RBQ: 'Repetitive questions: asking specific questions over and over'	*U*=362.0, *p*= .014
Answer is usually known to the questions asked *Included reports of the child being able to give the answer to the questions themselves if asked or providing the answer themselves within the question*	1.0	44.64	33.93	21.43		
Repetitive questions/ repetitive speech/ restricted conversation about the future *Children repeatedly asked or talked about what was coming next, what was going to happen, when things were going to happen or about events that were going to happen at some point in the future.*	0.77	54.55	39.39	6.06	RBQ: SUM of items; 'Repetitive questions', 'Repetitive phrases/ signing' and 'Restricted conversation'	*U*=249.0, *p*= .019
Temper outbursts usually follow the same course/ include the same behaviours	1.0	10.53	31.58	57.89		
When anxious repetitive speech/ repetitive self-injurious behaviour/ stereotypical behaviour can be shown *Child described as repeating the same phrase, showing stereotypical behaviour or self-injurious behaviour (including any behaviour that could potentially inflict tissue damage, even if tissue damage was not reported) when anxious.*	0.79	55.56	44.44	0.00		

Temper outbursts may occur when the child is perceived as manipulating the situation/ temper outbursts can be reinforced *Child showed an outburst because they wanted their own way or situations were described in which a temper outburst could be potentially reinforced i.e. by removing the aversive stimulus (e.g. a change) that triggered the outburst.*	1.0	42.86	46.43	14.29		
Anxiety may occur in social/ highly stimulatory situations *Anxiety was shown due to the people present in a situation, the noise, the visual stimulation or the smell*	1.0	84.62	15.38	0.00		
The individual does not appear to have control over their behavioural reaction (temper/ anxiety) *Children showed a behavioural reaction (temper outburst/ anxious reaction) that they were not able to control/ was automatic/ was something that they could not help/ was something that they had to do.*	1.0	8.82	26.47	64.71		
Warning is given of changes *When reported if and how advance warning of changes would affect the child's behaviour following the change*	0.83	50.00	31.58	18.42		
Repetitive activities *Repeatedly carrying out a particular activity, either at a particular time or at any time.*	1.0	78.69	21.31	0.00	RBQ: 'Lining up or arranging objects' CRI: 'Lines up objects into straight lines or symmetrical patterns'	(RBQ) U=387.5, p= .013 (CRI) U=410.0, p= .028
Stereotypical behaviour *Repetitive and seemingly purposeless movements of the hands/ parts of the body/ objects.*	1.0	61.54	26.92	11.54	RBQ: SUM of the three stereotypy items (object/ body/ hand)	U=337.5, p< .001

[2] Where operational definitions include expressions in quotation marks it is because the interview code was considered present when one of a number of specific words was used by participants.

Table 2. Shows the percentage of participants in each syndrome group who reported each interview code, and Chi-square and *p* values for comparisons between the groups. Cells are shaded where the difference between the groups is significant to a corrected level of $p < .003$

Behaviour code	**Percentage of participants who reported each interview code**		**Chi-Square values (df=1) and *p*-values for the comparison between the two syndrome groups**
	PWS	**FraX**	
Expect things to be as previously/ rigid thinking	73.9	12.1	$\chi^2=29.39$, $p< .001$
Change followed by negative emotional behaviour	87.0	78.8	$\chi^2= .93$, $p= .370$
Change followed by anger/ aggression	71.7	24.2	$\chi^2= 17.36$, $p< .001$
Change followed by temper outburst	89.1	39.4	$\chi^2=21.97$, $p< .001$
Change followed by anxiety	6.5	78.8	$\chi^2=43.19$, $p< .001$
Change followed by repetitive questions	78.3	75.8	$\chi^2= .068$, $p= .501$
Answer is usually known to the questions asked	78.3	57.6	$\chi^2= 3.89$, $p= .043$
Repetitive questions/ repetitive speech/ restricted conversation about the future	91.3	72.7	$\chi^2=4.82$, $p= .035$
Temper outbursts usually follow the same course/ include the same behaviours	41.3	0.0	$\chi^2=17.95$, $p< .001$
When anxious repetitive speech/ repetitive self-injurious behaviour/ stereotypical behaviour can be shown	0.0	54.5	$\chi^2=32.50$, $p< .001$
Temper outbursts may occur when the child is perceived as manipulating the situation/ temper outbursts can be reinforced	45.7	21.2	$\chi^2=5.02$, $p= .022$
Anxiety may occur in social/ highly stimulatory situations	0.0	39.4	$\chi^2=21.69$, $p< .001$
The individual does not appear to have control over their behavioural reaction (temper/ anxiety)	52.2	30.3	$\chi^2=3.75$, $p= .043$
Warning is given of changes	41.3	54.5	$\chi^2=1.35$, $p= .175$
Repetitive activities	67.4	87.9	$\chi^2=4.42$, $p= .031$
Stereotypical behaviour	28.3	39.4	$\chi^2=1.08$, $p= .213$